



ACTA RADIOLÓGICA PORTUGUESA

January | 2018
April | n° 1 | Volume 30



SPRMN

Sociedade Portuguesa
de Radiologia e Medicina Nuclear

EDITORIAL



Paulo Donato

One year has now passed since the online submission and editing system was implemented, a further step was taken to index the main databases of medical journals. Starting with this issue, all articles published in Acta Radiológica Portuguesa will be available in English.

Submission of the papers, as previously, may be done directly in English (without the Portuguese version), maintaining the possibility of submission in Portuguese. In this case, if the paper is accepted for publication, it will subsequently be translated into English by the Editorial Board of the Journal, according to the authors' corrections. Thus, when submitting in English, the paper will be available only in that language and when submitting it in Portuguese, the two versions will be published, one in Portuguese and one in English.

With this measure, we hope to maintain the national identity of the Journal and make international indexing easier.

In view of the interest shown by a large number of Radiologists present at the last SPRMN Conference, the person responsible for the updating program in Mammary Imaging, Dr. José Carlos Marques, was invited for the authorship of the article of opinion in the present issue of the Journal.

During the conference in Coimbra, the recent technological evolutions associated to the diagnosis of the medical image were also analyzed. Much has been said about Artificial Intelligence and its use in interpreting the data collected. About the growth potential for computing tools in imaging and in the role that the Radiologist will have in the future.

The advantages of Artificial Intelligence at the service of the Radiologist are a reality today. Nowadays, no one doubts the help constantly given in reading the imaging exams, namely in the diagnosis of pulmonary nodules, in the detection of pulmonary emboli, in the identification of fractures in conventional radiology, in the diagnosis of cerebral vascular accidents in urgent CT or even in the identification of breast nodules. However, the creation of databases of many billions of images with the most varied pathologies foresees the creation of self-learning systems, with a more comprehensive reading of the exams. The role of the Radiologist in the future depends on the meaning of this evolution. The future of Radiology will be subject to an in-depth analysis in future opinion articles.

At a time when many changes in imaging diagnosis are expected, there is also an increasing demand for Interventional Radiology. This demand regards not only the collection of tissue for a timely orientation of targeted therapy, but mainly the indication of urgent interventions with important prognostic implications and the development of more targeted and minimally invasive treatments, especially in the field of Oncology. Surely Radiology as a specialty will know how to adapt to these new requests.

We hope that, as in this issue, the Acta Radiológica will be considered for the publication of Radiology Intervention papers, with the demonstration of techniques that are done all over the country, some with great complexity and innovation, which need to be further disseminated at National level. I believe that this journal is the proper place for the scientific dissemination and empowerment of Intervention Radiology at National level.

A final word to mark the distinction made in the last ECR to the then President of SPRMN, Professor Filipe Caseiro Alves. He was awarded the gold medal of the European Society of Radiology in recognition of his important scientific work, especially in the area of gastrointestinal and abdominal radiology. It was a source of satisfaction and pride within SPRMN's direction and which was certainly generalized in the Portuguese Radiological community. It is an honorable individual recognition by the leading European Society of Radiologists, which is certainly related with the more global scientific development of the National Radiology.

Opinion Article / Artigo de Opinião

The Breast Radiology in Senology

A Radiologia Mamária na Senologia

José Carlos Marques



Instituto Português de Oncologia de Lisboa Francisco Gentil, Lisboa,
Portugal

In the last decade, technological advances and scientific knowledge about breast cancer have had a huge impact on all the subspecialties that make up the breast centers. In particular, Radiology has established itself as a central element in the multidisciplinary approach to breast pathology, intervening in all stages of breast cancer, including screening, diagnosis, staging and post-treatment follow-up.

This performance of Mammary Radiology was well reflected in the recent SPRMN conferences, dedicated to breast centers.

Using criteria for breast cancer mammographic screening based only on sex and age, as it has been so far the case, seems scarce in the light of current knowledge. It is important to identify high-risk groups that require different screening as well as factors that may increase cancer risk and decrease screening sensitivity, such as breast density. If for the high-risk groups of breast carcinoma, which include women with a heavy family history, a proven genetic mutation, or a history of treatment in the childhood with radiotherapy due to Hodgkin's lymphoma, there is consensus about the type of screening that should be done – annual breast MRI around the age of 25 - the same does not occur in the groups with high breast density (BI-RADS ACR-c and d), who represent about 40% of women at a traceable age. There is no established consensus as to the most appropriate complementary method to be used in the referred breast density patterns, namely ultrasound, MRI or tomosynthesis. Of these alternatives, since ultrasound is a labor-intensive and time-consuming examination and MRI is difficult to perform in such a large number of women, the advantage lies in tomosynthesis, which represents a recent advance in digital mammography, allowing the breast to be "sliced" in planes which are 1 mm thick.

Tomosynthesis allowed the elimination of false positives resulting from the overlapping of breast tissue and reduce false negatives by increasing the rate of detection of breast cancer in all mammary patterns, especially in the most heterogeneous and dense. The impact of tomosynthesis was so significant that its implementation was much faster than that of direct digital mammography. Its approval by the Food and Drug Administration (FDA) occurred in 2011 and today no one thinks of acquiring mammography equipment without tomosynthesis. There are several population screening programs under way with this technique, demonstrating exceptional results that reflect the significant increase in detection rate, as well as the reduction of false positives and consequently the reduction of the gauging rate.

On the other hand, the technological advance that allowed a synthesized 2D image from tomosynthesis, identical to the image of conventional mammography, with the same quality and without increasing radiation, also allowed an important dose reduction per examination. The synthesized 2D image constituted, therefore, a crucial stage in the process of tomosynthesis implementation.

MR is another indispensable modality in breast centers! In addition to its application in screening high-risk groups for breast cancer, the increased implementation in diagnostic settings has revealed high sensitivity and specificity, reflected in the amount of information provided when rigorous technical acquisition results in high quality examination.

One of the indications for breast MRI that has shown increasing adherence is the newly diagnosed breast cancer staging. Despite the discussion surrounding the topic, MRI has become a method that plays a pivotal role in surgical planning. The increased accuracy of MRI in the assessment of tumor extension and in the identification of in situ carcinoma component (CDIS) associated without mammographic translation, even in adipose patterns, has contributed to this performance. In addition, the rate of detection of multifocality and, to a lesser extent, bilaterality, as well as the evaluation of the ganglion chains.

The knowledge of the heterogeneity of breast cancer has added further indications for breast MRI at staging, namely in the molecular subtypes that are most frequently associated with multifocality, such as Her2 positive and luminal B tumors.

The number of situations with indication for neoadjuvant therapy has increased as a result of the advances in medical oncology with new and different therapeutic options, such as new antibodies for Her2 positive tumors. It also increases the rate of complete pathological responses obtained with neoadjuvant therapy, especially in triple negative, Her2 positive tumors and in tumors with high proliferative indexes (Ki67).

Current medicine is increasingly individualized and therapy is planned in a multidisciplinary environment based on the complete knowledge of the patient's characteristics and neoplasia. It is as important to know the molecular subtype of the lesion as to know if it is single or multiple, its extent and degree of involvement of the ganglion chains. Much of the information necessary for the complete picture of the neoplasm is provided by imaging techniques: mammography and tomosynthesis, ultrasound, and, especially, breast MRI.

Diagnosis of breast lesions using percutaneous biopsy techniques, guided by imaging methods, has been the common practice since the early 1990s and there is currently no room for surgical biopsy. Any surgery should be preceded by percutaneous diagnosis, since these are techniques of high precision and high concordance rate with the final histological diagnosis. This is why, in view of microcalcifications suspected of malignancy, it is mandatory that the biopsy be performed using a vacuum technique (BAV) in order to obtain a representative sample of the lesion, with a lower risk of disagreement with the final histology.

The advancement of biopsy techniques, the increased user experience and the greater knowledge about lesions of uncertain malignant potential or B3 lesions and their upgrading risk, have led to the increasing use of BAV not only for diagnostic purposes but also therapeutic ones. The current recommendations in B3 lesions, such as flat epithelial atypia, classic lobular neoplasia, papillary lesions and in the radiological scar are for the excision of these lesions by BAV and non-surgical, followed by imaging surveillance during 5 years.

In cases in where BAV reveals the diagnosis of CDIS, it is known that treatment may fail, with development of invasive carcinoma in 15 to 20% of the cases, and on the other hand there is a large proportion of treated patients that does not relapse during their life. These facts have reinforced the role of breast MRI in pre-surgical assessment of CDIS as a more sensitive and adequate technique for the evaluation of extension when compared to mammography, particularly in clinically significant tumors: major and of high

grade. They also raised the controversy of overtreatment, motivating international clinical trials to identify criteria that determine the progression to higher-grade CDIS and/or invasive carcinoma and, potentially, provide models based on the evidence of active surveillance as a future option of approach on low risk patients.

One area that is likely to change over the next few years is the follow-up after therapy. At present almost all international recommendations refer only to annual mammography. We know that there are subtypes that associate with a higher probability of early relapse, that there are dense patterns and surgical procedures that make it difficult to detect early the relapse with mammography. It is already possible to identify groups where the risk of relapse is greater and the follow-up should include MRI.

From the systemic point of view, the increasing importance attributed to the diagnosis of oligometastatic disease, due to the existing therapeutic options and with an apparent impact on survival, will also lead to a change in surveillance recommendations for the early detection of recurrence at a distance.

The challenges that Mammary Radiology poses to Medical Radiologists require dedication and knowledge that extend beyond the scope of Radiology. The scientific quality of the recent Conference, resulting from the sub-specialization of the participants and their integration in a multidisciplinary environment, was aimed at motivating and alerting the audience to the need and importance of the sub-specialization in Mammary Radiology.

Guidelines / Normas de Orientação

Multidisciplinary Clinical Protocol of Management of Hypersensitivity* Reactions to Contrast Media in Radiology

Protocolo Clínico Multidisciplinar de Abordagem das Reações Agudas de Hipersensibilidade a Meios de Contraste em Imagiologia

Inês Rolla¹, Cristina Lopes², Ernestina Gomes³, M. Catarina Tavares⁴

¹ Interna de Formação Específica de Radiologia

² Assistente Hospitalar Graduado de Imunoalergologia

³ Assistente Hospitalar Graduado de Medicina Intensiva

⁴ Assistente Hospitalar Graduado de Radiologia Hospital Pedro Hispano, Matosinhos Portugal

Address

M. Catarina Tavares
Radiology Department
Pedro Hispano Hospital
Matosinhos
Portugal
email: mcatarinatavares@gmail.com

Abstract

Acute hypersensitivity reactions to contrast media (AHRC) are infrequent, usually mild but potentially fatal. Although there are recommendations from Radiology, Intensive Care and Allergology fields regarding its management, a clinical multidisciplinary protocol that integrates these complementary approaches is missing.

We aimed to elaborate a protocol that includes adverse reactions definition and classification; identification of risk factors and management of patients with AHRC.

A non systematic revision of national and international guidelines was made regarding the management of hypersensitivity reactions, in order to elaborate a clinical consensus protocol to be used in different medical fields (Radiology, Allergology and Intensive Care).

Non-renal adverse contrast reactions can be classified in chemotoxic (related to contrast chemical properties) and hypersensitivity reactions (with involvement of immunological mediators IgE and non IgE mediated); and in mild, moderate and severe (regarding severity), with different therapeutic approaches. Identified risk factors are previous contrast media reaction and asthma. Pre-medication decreases the probability of symptoms but does not exclude the possibility of a severe reaction. Patients suspected of AHRC should be observed in an allergy clinic to confirm the diagnosis and find an alternative contrast media. Clinical alert record as well as the notification of reaction to surveillance system of adverse drug reactions should be performed.

This clinical expert's protocol consensus based on national and international guidelines aims to be a valuable practical tool in the management of patients that need contrast media during a radiologic exam.

Keywords

Contrast media; Hypersensitivity; Adverse reaction; Radiology.

Resumo

As reações agudas de hipersensibilidade a meios de contraste (RAHC) em imagiologia são consideradas pouco frequentes; são habitualmente ligeiras, mas podem ser potencialmente fatais. Apesar de existirem recomendações nacionais e internacionais sobre a sua abordagem, não existe um protocolo clínico que integre a visão de todas as especialidades envolvidas (Imagiologia, Imunoalergologia e Medicina Intensiva).

Pretendeu-se estabelecer um protocolo prático de utilização transversal e adequada à realidade hospitalar, que incluía a definição das reações adversas e sua classificação, a identificação de factores de risco para a ocorrência de reacção de hipersensibilidade, a abordagem diagnóstica e terapêutica e de orientação dos doentes com RAHC.

Realizou-se uma revisão não sistemática das recomendações nacionais e internacionais acerca da abordagem das RAHC, publicadas pelas sociedades científicas de Imagiologia, Imunoalergologia e Medicina Intensiva e o protocolo foi elaborado por consenso de peritos destas especialidades.

As reações adversas não renais subdividem-se em reações quimiotóxicas (relacionadas com as características de cada contraste) e reações de hipersensibilidade (com envolvimento de mediadores imunológicos de forma IgE e não IgE mediada); podem ser classificadas em ligeiras, moderadas e graves com abordagens terapêuticas específicas. Constituem os principais factores de risco para a sua ocorrência, a reacção prévia a meio de contraste e a asma brônquica. A pré-medicação diminui a probabilidade de ocorrência de sintomas, mas não exclui a possibilidade de reacção grave. Os doentes com suspeita de reacção de hipersensibilidade devem ser encaminhados para a consulta de Imunoalergologia para diagnóstico definitivo, orientação para redução de risco e escolha de meio de contraste alternativo. A inserção de alertas clínicos, assim como a notificação deve ser assegurada.

Este protocolo clínico de consenso de peritos, baseado nas indicações nacionais e internacionais atuais, pretende ser uma ferramenta passível de utilização na prática clínica aquando da abordagem do paciente a realizar exames imagiológicos que necessitem de meio de contraste.

Palavras-chave

Meios de contraste; Hipersensibilidade; Reacção adversa; Imagiologia.

*Authors' note: In this document, and following the SEAIC designation proposal⁷, acute non-renal adverse reactions, allergic and allergic-like, are referred to as acute contrast media hypersensitivity reactions in imaging.

1. Objectives

- To address the definition of acute hypersensitivity reaction to contrast media and the differential diagnosis with chemotoxic reactions;
- To determine risk factors for acute hypersensitivity reactions;
- To establish laboratory procedures to be performed in case of suspected allergic reaction;
- To standardize criteria for clinical diagnosis and treatment of acute hypersensitivity reactions; and
- To define orientation criteria for the allergology appointment.

Delayed adverse reactions (from one hour to several days after administration of contrast media) and nephrotoxic reactions are outside the scope of this protocol.

This protocol should be applied to all patients undergoing a radiologic study with administration of contrast media as part of the best practices associated with the use of contrast media in any radiology department.

Various contrast media and various routes of administration are used in imaging; by the frequency and clinical significance of associated acute adverse reactions, this protocol refers only to iodinated contrast media and gadolinium-based ones when administered intravenously or intra-arterially.

2. Definitions

An acute non-nephrotoxic adverse reaction to a contrast media is a noxious and unintentional reaction that occurs up to one hour after the administration of the contrast media and does not result from the nephrotoxic effect.

These reactions can be observed with iodinated contrast media in procedures such as Angiography, Urography and Computed Tomography and with gadolinium contrast agents in Magnetic Resonance Imaging being, however, more frequent with iodinated contrast media. In terms of severity, the majority of reactions are mild, whereas life-threatening situations are rare.

Acute non-nephrotoxic adverse reactions may be subdivided into chemotoxic reactions and hypersensitivity reactions.

Chemotoxic reactions:

- are related to the chemical properties of the contrasts administered, and generally depend on the dose and on the infusion rate;
- are usually transient and self limited, but can be serious. They frequently include heat sensation, flushing, nausea and vomiting;
- vagal vase reactions are a less frequent type of chemotoxic reactions characterized by hypotension and bradycardia;
- cardiovascular adverse effects should also be considered in the context of chemotoxic reactions, such as arrhythmias, cardiac contractility depression, hypertensive crisis and cardiogenic pulmonary edema. In cases of severe hypotension, loss of consciousness, cardiorespiratory arrest, angina, or seizure may occur. These effects are very rare;
- the occurrence of these reactions does not prevent the administration of contrast media in future imaging studies.

Hypersensitivity reactions:

- are idiosyncratic and unpredictable, and may occur in response to administration of minimal amounts of contrast media
- can be allergic, mediated by IgE / T lymphocytes or allergic-like mediated by non-specific mechanisms of release of vasoactive mediators (direct endothelial membrane effect, complement system activation or direct formation of bradykinins).

3. Classification of adverse reactions according to severity

The AHRC classification can be made according to clinical severity, in mild, moderate and severe reactions (Table 1).

In cases of severe cardiovascular reactions, such as pulmonary edema, significant hemodynamic instability and cardiorespiratory arrest, although these reactions may result from a chemotoxic reaction, an allergic etiology may be assumed for future treatment and referral of the patient, especially if signs and symptoms of skin and mucosa involvement coexist.

Table 1 - Classification of adverse reactions according to severity

Hypersensitivity reactions	Chemotoxic reactions
Mild	
Limited urticaria / pruritus Limited skin edema Scratched throat Nasal congestion Sneezing / conjunctivitis / rhinorrhea	Limited vomiting and nausea "Flushing" / heat sensation Transient tremors Headache / dizziness Anxiety Taste alteration
Moderate	
Generalized urticaria / pruritus Diffuse erythema with stable vital signs Facial edema without dyspnea Wheezing / slight bronchospasm	Vomiting and severe nausea Hypertensive urgency Isolated chest pain Vasovagal reaction (which requires treatment and responds to treatment)
Severe	
Facial or generalized edema with dyspnea Diffuse erythema with hypotension Laryngeal edema with stridor and/ or hypoxia Wheezing / bronchospasm with hypoxia Non-cardiogenic pulmonary edema Anaphylactic shock	Resistant vasovagal reaction Arrhythmias Convulsions Hypertensive emergency Cardiogenic pulmonary edema

4. Classification of contrast media

Iodinated contrast media (ICM) are iodine salts whose chemical structure includes a benzene ring with at least 3 iodine atoms which is responsible for the radio-opacity.

The ICM can be classified according to iodine molecule loading (ionic and nonionic), molecular structure (monomeric and dimeric) and osmolality (hyperosmolar, low osmolality and iso-osmolality). Contrast osmolality is the most frequently associated factor with hypersensitivity

reactions and reactions are more frequent with those with high osmolality.

Most ICM are classified as:

- monomeric ionic - have the highest osmolality, are essentially used in extravascular procedures such as cystograms;
- dimeric ions - the only example is ioxaglate and is low in osmolality;
- monomeric nonionic - are considered second generation, have low osmolality - examples: iohexol, iopamidol, ioversol, iopromide, ioxylan, iomeprol;
- dimeric non-ionic - example: iodixanol - is associated with equal or lesser number of hypersensitivity reactions than those of low osmolality.

The contrast media used in magnetic resonance are gadolinium complexes that can be classified as ionic or nonionic and according to their structure in linear or macrocyclic.

5. Prevalence

With low osmolality ICM, the prevalence of acute hypersensitivity reactions is very low (0.2% to 0.7%) and severe reactions are even rarer (0.04%).

With gadolinium-based contrast media the prevalence of acute hypersensitivity reactions is much lower than with iodinated contrast (ranging from 0.24% to 0.07%), and severe allergic reactions are extremely rare (with prevalence of 0.01 to 0.001%).

6. Risk factors for hypersensitivity reaction

Two types of risk factors can be considered

a) Relative to the contrast media

Reactions are more frequent with high osmolality contrast media, however the prevalence of serious reactions seems to be independent of the degree of osmolality.

b) Regarding the patient

The main risk factors for contrast media hypersensitivity reactions are:

- Previous history of AHRC in an imaging study with contrast media administration.
- Unstable asthma

Some authors consider that previous history of bronchial asthma, drug allergy, food allergy, female gender may be a relative risk factor, however, these do not prevent the administration of contrast media nor do they determine the need for premedication.

It should be noted that the occurrence of a chemotoxic reaction in a previous exam with intravenous contrast media is not a risk factor for the occurrence of hypersensitivity reaction, therefore there is no need to pre-medicate these patients.

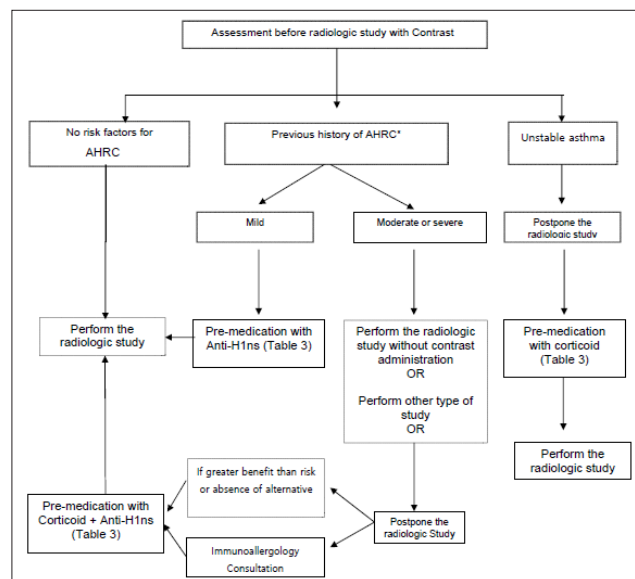
The approach of the patient submitted to a radiologic study with contrast media is explained in Figure 1.

7. Attitudes to be taken in patients with risk factors

a) General measures

- There should be reciprocal information and coordination between the team of technicians and nurses, and the neuro / radiologist;

Figure 1 - Approach of the patient who will be examined with contrast medium



* To ICM if a CT or angiography or urography is indicated; to gadolinium-based contrast media if a MRI is indicated.

Anti-H1 ns- antihistamine H1non sedative

- The patient should remain in the imaging service, in the area of the CT / MRI, with venous access, for 30 min;
- If you give an antihistamine with a torpor effect (eg clemastine IV) to a patient, he/she should not drive or work with machines within 6 hours or as long as the symptoms of drowsiness remain.

b) if there is a history of mild hypersensitivity reaction

- The examination must be performed;
- Intravenous contrast can be performed, if important for the diagnosis;
- If the CM to be administered is identical to that associated with previous adverse reaction, consider non-sedating antihistamine 30 minutes before the test (Tables 2 and 3)

c) If history of moderate to severe hypersensitivity reaction

- Consider performing the radiologic study without contrast media;
- Consider carrying out another type of study;
- Postpone examination and:
 - If possible, refer the patient to an allergology consult in order to carry out an allergy study and establish a diagnosis and specific guidelines;
 - According to the benefit, especially in the absence of an alternative, perform the imaging study with contrast media after “premedication” measures, according to the clinical situation - Corticoid + Anti-H1ns (Tables 2 and 3).
- In this situation, consider using another active pharmacological principle of contrast media, different from the one used in the previous study in which the hypersensitivity reaction occurred.

d) If unstable asthma

In case the patient reports medical diagnosis of asthma and presents with uncontrolled symptoms such as wheezing or bronchospasm, dyspnea and low effort tolerance:

- Consider performing the radiologic study without contrast media;

- If it is necessary to perform a radiologic study with CM administration, it must be postponed and have premedication programmed - corticoid - in this particular case it is important to perform the corticoid as an anti-edematous (Tables 2 and 3).

e) If history of allergic reaction to other allergens

Follow the general indications specified above; this situation, by itself, does not require premedication.

Table 2 - Pre-medication scheme according to risk factors

Pre-medication scheme
<p>Previous history of mild hypersensitivity reaction to contrast</p> <p>Non-sedative anti-H1 2nd generation, orally (PO), 30 min before contrast administration (Table 3)</p>
<p>Previous history of moderate to severe hypersensitivity reaction to contrast and examination cannot be postponed waiting for allergology appointment</p> <p>a) Adults and >12 year-old children Prednisolone 50mg 13, 7 and 1 hour before PO or methylprednisolone 32mg PO 12h and 2h before PLUS Non-sedative anti-H1 2nd generation PO 2 h prior to administration of contrast media</p> <p>If the patient cannot take oral medication choose for i.v pre-medication: Ex: Hydrocortisone 200mg iv, 12h and 2h before contrast + Clemastine 2mg 2h prior to administration of contrast media</p> <p>If situations not programmed arise (patients in the ER): Ex: Hydrocortisone 200mg and every 4h / 6h, until the radiologic study + clemastine 2mg ev 1 hour before contrast.</p> <p>b) ≥ 6 year-old children Prednisolone 0.5 mg / kg PO 13, 7 and 1 h before or methylprednisolone 1 mg / kg PO 12 h and 2 h before PLUS Non-sedative anti-H1 2nd generation per os 2 h prior to administration of contrast media</p> <p>c) < 6 year-old children Ex: Betamethasone 0.03 mg / kg PO (in general it corresponds to 2 drops per kg); 12 and 2 hours prior to administration of contrast agent PLUS Non-sedative anti-H1 2nd generation PO 2 h prior to administration of contrast media</p>
<p>Unstable asthma</p> <p>a) Adults and >12 years-old children ex: prednisolone 0,5mg PO, 12h and 2h prior to contrast media administration.</p> <p>b) ≥ 6 years-old children ex: prednisolone 0,5mg/Kg (max 50mg), PO, 12h and 2h prior to contrast media administration.</p> <p>c) < 6 years-old children ex: betamethasone 0,03 mg/Kg PO (in general it corresponds to 2 drops per Kg); 12h and 2h prior to contrast media administration.</p>

8. Considerations for chemotoxic reactions

In order to minimize chemotoxic reactions in contrast medium in imaging, we must:

- Administer contrast in imaging study only if there is a diagnosis benefit;
- Comply with the manufacturer’s handling and packaging instructions;

Table 3 - Drugs that can be used as pre-medication

Name	Formulation	Children dose	Adults dose
Antihistamine			
Fexofenadine	S,C	6-12 year-old: 30 mg >12 year-old:120-180 mg 1x/day	120-180 mg
Cetirizine	S,C	2 - 5 year-old: 2,5 mg 6 - 11 year-old: 5 mg >12 year-old: 10 mg/ day	10 mg
Loratadine	S,C	2 - 11 anos: ≤ 30 kg: 5 mg > 30 kg: 10 mg >12 year-old, > 30 kg: 10 mg	10 mg
Levocetirizine	S,C	2 - 5 year-old: 1,25 mg 6 - 11 year-old: 5 mg > 12 year-old: 5 mg	5 mg
Desloratadine	S,C	1 - 5 year-old: 1,25 mg 6 - 11 year-old: 2,5 mg > 12 year-old: 5 mg	5 mg
Ebastine	S,C	2 - 5 year-old: 2,5 mg/ day 6 - 11 year-old: 5 mg/ day >12 year-old: 10,0 mg/ day	10-20 mg
Rupatadine	S,C	2 - 11 year-old, 10 - 25 kg: 2,5 mg ≥ 25 kg: 5,0 mg > 12 year-old: 10 mg	10 mg
Bilastine	C	> 12 year-old: 20 mg/ day	20 mg
Clemastine	IM, IV	0,025 mg/Kg	2 mg
Corticoids			
Prednisolone	C, IV	0,5 mg/Kg PO 0,5 mg/Kg IV	50 mg PO 50 mg IV
Metilprednisolone	C, IV	1 mg/Kg PO 0,5 mg/Kg IV	32 mg PO 40 mg IV
Hydrocortisone	IV	4 mg/Kg	200 mg
Betametason	S	< 6 year-old: Betamethasone 0,03 mg/Kg PO (corresponding in general to 2 drops per Kg);	
Deflazacort	S,C	< 6 year-old 1 mg /Kg(= 1 drop per Kg)	30 mg

S-Oral solution C-pill A-years PO-orally IV-intravenous

- In the case of ICM, use iso-osmolar or low osmolarity contrast agents;
- Administer the minimum required dose/volume and the lowest flow rate possible, according to the clinical indication for the radiologic study;
- In the case of ICM, to calculate volume and flow rate, the patient’s profile should also be taken into consideration, namely the presence of cardiac pathology and the risk of water overload;
- Realize that anxiety, on its own, may be responsible for the occurrence of symptomatology identical to a true adverse reaction to contrast media.

9. Treatment of non-renal acute adverse reactions to contrast media

Any patient may develop, in any contrast-enhanced radiologic study, an acute adverse reaction, which should be promptly identified and treated. The patient should be guided in order to obtain a clinical or clinical-laboratory diagnosis, with implications in future radiologic studies.

The approach of acute non-renal adverse reactions to contrast media is summarized in Figure 2.

There are a number of universal measures that should be taken in all adverse reactions, including suspension of contrast administration, oxygen mask and monitoring:

- Elimination of the triggering factor is essential, meaning in this case the interruption of contrast media infusion.
- Every patient should be placed in a comfortable position. Patients may prefer to be seated as it will ease their breathing. Dorsal decubitus, with elevation of the lower limbs, is useful in hypotensive patients and when we think a vagal reaction is occurring. If you feel that the patient is going to faint, he/she should not sit or stand up. The patient should be placed where he can easily take off the clothes in order to observe the progression of skin lesions.
- Monitoring should always include at least respiratory rate, pulse oximetry, noninvasive blood pressure, and heart rate.
- Oxygen must be placed by facial mask (high concentration mask) until the clinical situation is reevaluated.

In the event of a mild reaction, only the universal measures should be taken and if the symptoms are resolved, the patient's surveillance must be kept in the radiology department for at least 30 minutes after the resolution of the symptoms.

If the suspicion is mild hypersensitivity reaction, a second generation non-sedative antihistamine drug may be used. In the event of a moderate or severe reaction, the Medical Emergency Team (MET) must be immediately activated. If a hypersensitivity reaction is suspected, the on duty imaging physician should immediately administer Adrenalin IM (see Table 4). Until the arrival of the MET, the professionals present must take the Basic Life Support measures appropriate to the clinical case.

9.1. Treatment of chemotoxic reactions

The treatment of these reactions is symptomatic and directed to the type of reaction.

The majority of reactions are mild; it is advisable to keep patient surveillance in the imaging department until 30 minutes after the resolution of symptoms.

If the reaction is moderate or severe, the MET should be involved to do intensive treatment. In this case the surveillance should be extended until 8 hours, in an inpatient regime.

9.2. Treatment of hypersensitivity reactions

9.2.1 Mild reactions

In the event of a mild reaction, the universal measures, monitoring and surveillance described above must be taken.

The indicated antihistaminic drugs are non-sedative (Table 3) or in the case of impossibility of oral use, for example

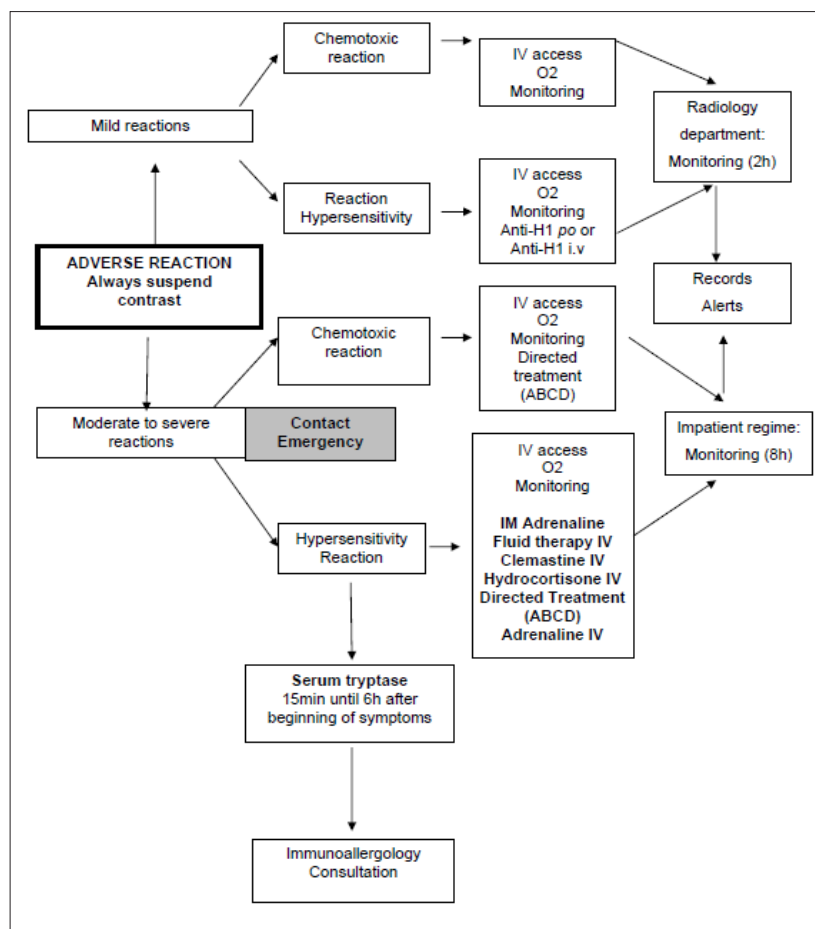


Figure 2 – Treatment of hypersensitivity reactions to contrast media

Clemastine (the adult dose is 2 mg IV or IM, in children the correct dose is 0.025 mg / kg / IM or EV dose (up to a maximum of 2 mg) may be used, see table 4.

9.2.2 Moderate or severe

A moderate or severe AHRC is likely to be present when a patient develops sudden and unexpected illness (usually within minutes after administration of contrast media), with rapidly progressing skin changes and potentially fatal airway and / or respiratory and / or circulatory system compromise. The allergic reaction, being a specific and potentially fatal type of adverse reaction, has a specific approach. This approach should be appropriate to the clinical severity of the illness. This acting algorithm is based on the recommendations of the European Resuscitation Council (ERC).

ABCDE approach

The ABCDE methodology of recognition and approach of any critical patient is the one that should be used in these situations.

Airway (A - airway) problems may correspond to edema of the airways, and may be manifested as hoarseness or stridor and may progress to complete obstruction and cardio-respiratory arrest. Airway obstruction can occur quickly. The warning signs are edema of the tongue and lips and hoarseness. Early involvement of an airway expert (experienced anesthesiologist, intensivist) is mandatory in the treatment of these patients. It is necessary to consider early tracheal intubation; any delay can make intubation extremely difficult. A surgical airway may be necessary if tracheal intubation is not possible.

Breathing problems (B - breathing) can lead to dyspnea; wheezing, whistling; cyanosis; and confusion, frequently related to the presence of bronchoconstriction and respiratory failure.

Circulatory problems (C - circulation) can range from hypotension to cardiac arrest. Circulatory problems (often referred to as anaphylactic shock) may be caused by direct myocardial depression or vasodilatation and increased capillary permeability.

Airway, Breathing, and Circulation problems referred above may alter the patient's neurologic status (**D - disability**) due to decreased cerebral perfusion. There may be confusion, agitation and loss of consciousness.

Adrenalin

It should be noted that if there is suspicion of moderate or severe hypersensitivity reaction, the drug of choice is Adrenalin.

There are no absolute contraindications for treatment with intramuscular adrenalin in an anaphylaxis situation. Adverse effects are very rare when the correct doses are injected intramuscularly (IM). Subcutaneous or inhalation routes are not recommended.

In adults and children over 12 years of age, administer an initial dose of 0.5 mg IM adrenalin (0.5 ml ampoule 1 mg / ml (1: 1000 adrenalin) = 0.5 mg = 500 mcg). Additional doses may be administered at intervals of about 5 min, depending on the patient's response, up to 3 administrations.

In children 6 to 12 years of age the dose of adrenalin is 0.3 mg IM (0.3 ml ampoule 1 mg / ml (1: 1000 adrenalin) = 0.3 mg = 300 mcg). In children less than 6 years of age the dose of adrenalin is 0.15 mg IM (0.15 ml ampoule 1 mg / ml (1: 1000 adrenalin) = 0.15 mg = 150 mcg) (Table 4).

Table 4 - Drugs used in the treatment of hypersensitivity reactions

ADRENALINE (always administered IM - dose by weight - 0.01 mg / kg / dose)
Adult and >12 year-old child - administer an initial dose of 0.5 mg IM adrenalin (0.5 ml 1 ampoule 1 mg/ml = 0.5 mg = 500 mcg). Additional doses may be given at intervals of about 5 min up to 3 doses.
6 to 12 year-old Children - administer an initial dose of 0.3 mg of IM adrenalin (0.3 ml of 1 ampoule of 1 mg/ml = 0.3 mg = 300 mcg).
<6 year-old Children - administer an initial dose of 0.15 mg IM Adrenalin (0.15 ml 1 vial of 1 mg/ml = 0.15 mg = 150 mcg).
Adrenaline IV (only in case of lack of response to IM adrenaline and by a doctor with experience in its use)
(dilute one ampoule of adrenaline (1mg/ml) up to 100ml of saline solution - a concentration of 10ug / ml)
Adult: Bolus of 20 to 50 ug repeated every 1-2 minutes if necessary; perfusion of 0.1-1 ug / kg / min, the same dilution can be used Pediatrics: continuous infusion 0.1-1 ug / kg / min. Bolus IV not applicable.
FLUIDS
Adult - fast fluid bolus IV (500-1000 ml crystalloid), if necessary administer additional doses.
Children - fluid bolus IV (20 ml/kg of crystalloids).
OTHER DRUGS
Clemastine – 0,025 mg/kg/dose EV/IM (max - 2mg)
Hydrocortisone – adult - 200 mg; children - 4 mg /kg
Glucagon (resistance to adrenaline in beta-blocked patients) - bolus of 30 µg/kg / IV dose up to a maximum of 1 mg (may be repeated every 5 minutes) Continuous perfusion 5-15ug/min (dilute one ampoule to 100ml of saline solution (10ug/ml))

The best place for IM injection is the anterolateral zone of the middle third of the thigh. The needle must be long enough to ensure that adrenalin is injected into the muscle (21G green color, 4 cm, also known as an intramuscular needle).

IV adrenalin should only be administered by professionals experienced in the use and titration of vasopressors in clinical practice (eg: anesthesiologists or intensivists). In patients with spontaneous circulation, intravenous adrenalin may cause potentially fatal hypertension, tachycardia, arrhythmias, and myocardial ischemia. Do not administer the 1: 1,000 IV adrenalin solution without proper dilution (dilute 1 mg to 100 ml saline, you can use this dilution for bolus or for infusion). Titrate adrenalin IV in bolus of 20 to 50 mcg (2 to 5 ml of the proposed dilution) depending on the response. In children, do not titrate Adrenalin IV in bolus. If repeated doses are required, establish IV infusion of adrenalin using the existing tables (1 to 10 mcg / min is the usual dose which corresponds to 6 to 60 ml/h of the previous dilution, titrating for the intended effect).

Adrenaline is the first-line vasopressor for the treatment of allergic reactions. Ponder other vasopressors and inotropes (noradrenaline, vasopressin, terlipressin) when the initial resuscitation with adrenalin and fluid is not successful. These drugs should only be used in specialized settings (eg: in intensive care units) where there is experience in their

use. Glucagon may be useful in treating allergic reactions in patients taking beta-blockers.

Fluid therapy

Timely fluid therapy is one of the determining factors to avoid and correct hypotension / distributive shock that may lead to cardiorespiratory arrest.

Rapid fluid bolus IV (500-1000 ml adult crystalloid) should be administered, monitoring the response and if necessary administering additional doses. In children, a bolus of crystalloid fluids of 20 ml/ kg is recommended. In this context, Crystalloid are preferable as they are isotonic and glucose-free use (such as Poly® or Plasma-Lyte®).

Other drugs

The administration of antihistamines, although reducing the symptoms is not life saving and should never delay the administration of intramuscular adrenaline. The suggested antihistamine drug is Clemastine. The adult dose is 2 mg I.V or I.M. In the child the correct dose is 0.025mg/kg/dose I.M. or E.V (up to a maximum of 2mg).

There is little evidence to support the routine use of H2 antihistamines (eg ranitidine, cimetidine) for the treatment of these conditions.

The administration of corticosteroids is mainly for the prevention of biphasic reactions (resurgence of allergic reaction after 6 to 8 h) and it is neither life saving and should never delay the administration of intramuscular adrenalin. There is little evidence as to the ideal dose of corticosteroids; for example, the hydrocortisone suggested dose in adults is 200 mg IV and in child 4 mg/kg IV slow.

Cardiorespiratory arrest

In the event of a cardio-respiratory arrest, advanced life support measures should be immediately initiated, in accordance with the latest ERC guidelines.

10. Laboratory procedures

The diagnosis of hypersensitivity reactions is clinical. There are several differential diagnoses that should be considered according to the severity of the clinical manifestations, and excluded when necessary, never delaying the treatment intervention.

In all moderate to severe AHRC, a blood sample should be taken for quantification of serum tryptase levels. When? as early as possible between 15 minutes to 6 hours after the onset of symptoms.

In all moderate to severe AHRC, an allergology appointment is required. The increase of serum triptase supports the diagnosis of IgE-mediated hypersensitivity, but, if normal, an anaphylactic reaction must not be excluded. In case of increase in the serum triptase value, a new sample should be taken after the resolution of the symptoms, during the patient's follow-up. Other complementary studies will be performed during follow-up.

11. Discharge and follow-up

Patients with moderate to severe hypersensitivity reactions should be treated and observed for at least 8 hours in a facility that is capable of treating these conditions. The exact incidence of biphasic reactions that could warrant prolonged surveillance is unknown. There is no reliable way to predict who will undergo a biphasic reaction. Thus, it is important that discharge decisions are made for each individual patient by an experienced physician. There is no indication for outpatient maintenance if all symptoms are resolved.

Before discharge, all patients should:

- Be informed about the nature of the reaction and the need to continue clinical research. The patient should also be informed that he/she should avoid the contrast media until he/she goes to the allergology appointment.
- Get clear instructions to return to the hospital if symptoms resurface.
- Be referred to a specialist in allergology.

11.1 Referral to an allergology medical appointment

The following patients should be referred to an allergology appointment:

- Patients with increased risk of contrast media hypersensitivity reaction (suspicion of moderate or severe reaction prior to contrast product). Referral by the imaging physician or the physician requesting the radiologic study.
- Patients who had a moderate or severe AHRC.

12. Records

12.1 Clinical records

Whenever an acute non-nephrotoxic adverse reaction occurs to the contrast media a clinical record should be made. This record should contain a description of the clinical manifestations, therapy performed and time elapsed until resolution of symptoms.

12.2 Pharmacovigilance registration

The adverse reaction should also be recorded on the "National Registry" (eg, INFARMED in Portugal).

13. Alerts

The insertion of the hypersensitivity reaction information into the patient's clinical / electronic process is fundamental to prevent future administrations and reactions.

Received / Recebido 19/11/2017
Acceptance / Aceite 03/01/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

References

1. World Allergy Organization Guidelines for the Assessment and Management of Anaphylaxis, F. E. R. Simons.
2. Norma da DGS N.º 014/2012 – Anafilaxia: Abordagem Clínica.
3. Norma da DGS N.º 004/2012 – Anafilaxia: Registo e Encaminhamento.
4. ACR Manual on Contrast Media Version 10.3 2017 ACR Committee on Drugs and Contrast Media.
5. European Resuscitation Council Guidelines for Resuscitation 2015: Section 4. Cardiac arrest in special circumstances. Anatolij Truhlar et al. Resuscitation, October 2015, Pages 148 – 201.
6. ESUR Guidelines on Contrast Media, version 9.0, 2016, European Society of Urogenital Radiology.
7. Clinical Practice Guidelines for Diagnosis and Management of Hypersensitivity Reactions to Contrast Media J Investig Allergol Clin Immunol. 2016; 26:144-55.

Endovascular Treatment of Splanchnic Pseudoaneurysms Following Blunt Solid Organ Injuries in Children

Tratamento Endovascular de Pseudo-Aneurismas após Trauma Abdominal Fechado em Crianças

João André Oliveira¹, Nuno Vasco Costa², Tiago Bilhim², Filipe Veloso Gomes², Élia Coimbra²

¹ Serviço de Radiologia, Hospital Geral de Santo António, Centro Hospitalar do Porto, Portugal
² Serviço de Radiologia, Hospital Curry Cabral, Centro Hospitalar de Lisboa Central, Portugal

Address

João André Oliveira
Rua das Artes Gráficas, 67-71
4100-092 Porto, Portugal
e.mail: joao_a_oliveira@hotmail.com

Abstract

Non operative management is currently the standard treatment of blunt abdominal solid organ injuries grades I-IV (American Association for the Surgery of Trauma's organ injury scale) in children. Even though post-traumatic splanchnic pseudoaneurysms are an infrequent complication, they may potentially lead to life-threatening intra-peritoneal or retroperitoneal bleeding. In adults, the relationship between failure of conservative management in abdominal trauma patients and delayed rupture of a pseudoaneurysm identified in follow-up imaging is well established, as is the capability of selective angioembolization to decrease non operative management failure rate. In the pediatric population, the clinical and prognostic significance of splanchnic pseudoaneurysms remains controversial and, currently, there are no high-level evidence-based guidelines on its management. The authors of this paper present 3 cases of post-traumatic abdominal pseudoaneurysms in children which were identified in imaging exams after conservative management, successfully treated by selective embolization, and a review of the literature regarding this subject is also presented. Although prospective randomized-controlled trials are needed to better define the incidence, natural history and optimal management of abdominal PAs in pediatric blunt abdominal trauma, we believe that selective angioembolization provides a safe and effective therapy for its treatment and should be considered as part of the multidisciplinary trauma management protocol in children.

Keywords

Blunt abdominal trauma; Children;
Pseudoaneurysm; Interventional radiology;
Angioembolização.

Resumo

A abordagem conservadora é atualmente o tratamento de primeira linha de lesões traumáticas de órgãos sólidos intra-abdominais de grau I-IV (escala da Associação Americana de Cirurgia de Trauma) em idade pediátrica. Embora pouco frequentes, os pseudoaneurismas esplâncnicos pós-traumáticos podem potencialmente originar hemorragia intraperitoneal ou retroperitoneal catastrófica. Na população adulta, a rotura tardia de pseudoaneurismas é uma das causas do insucesso da abordagem conservadora. A angioembolização seletiva destes pseudoaneurismas contribui para a diminuição da taxa de insucesso desta abordagem. Na população pediátrica, a relevância clínica e prognóstica dos pseudoaneurismas esplâncnicos não se encontra ainda clarificada, e atualmente, não há guidelines de alto nível de evidência para a sua abordagem terapêutica. Os autores deste artigo apresentam 3 casos de pseudoaneurismas abdominais pós-traumáticos em crianças que foram identificados em exames de imagem após tratamento conservador, tratados com sucesso por embolização seletiva, e apresentam uma revisão da literatura no que concerne a este tema. Apesar de estudos prospetivos randomizados serem ainda necessários para melhor definir a incidência, a história natural e a abordagem terapêutica dos pseudoaneurismas abdominais no trauma pediátrico, os autores defendem que a angioembolização seletiva representa uma abordagem segura e efetiva no tratamento desta entidade clínica e que deve ser integrada no protocolo da abordagem multidisciplinar do trauma pediátrico.

Palavras-chave

Trauma abdominal; Crianças; Pseudo-aneurisma;
Radiologia de intervenção; Embolização.

Introduction

Abdominal post-traumatic pseudoaneurysms (PAs) are a relatively rare complication following blunt trauma, with disruption of the arterial wall and formation of a blood-perfused sac contained by the media/adventitia layers or in some cases by the soft-tissues encircling the injured vessel. Due to continuous high arterial pressure feeding the PA, there is a potential life-threatening risk of intra-abdominal or retroperitoneal bleeding.

In adults, the relationship between failure of conservative management in abdominal trauma patients and delayed rupture of a PA identified in follow-up imaging – namely in the liver, kidney and spleen territories - is well established, and most trauma centers perform prophylactic angiographic embolization of abdominal PA in hemodynamically stable patients.

In the pediatric population, clinical and prognostic significance of traumatic abdominal PAs is not well defined. For instance, small splenic artery PAs are thought to frequently resolve spontaneously with thrombosis of the

aneurismatic sac.¹⁻⁷ On the other hand, hepatic artery PAs have an up to 80% risk of rupture with potential catastrophic bleeding, and the incidence of spontaneous thrombosis is low.^{8,9} Thus, controversy remains regarding the management of PAs identified on follow-up imaging, and strategies are often influenced by the clinicians' preferences and expertise in each center.

The authors present 3 cases of post-traumatic abdominal PAs in children successfully treated with angioembolization at our institution reviewing the literature related with this matter.

Case 1

A 6-year-old boy came to our institution due to upper abdominal trauma after hitting a pool handrail. In the initial assessment, he was found to be conscious, tachycardic (119 BPM) but hemodynamically stable, with pain referred to the left upper quadrant. His complete blood count (CBC) showed a decreased hemoglobin level (10,4g/dL). A contrast-enhanced CT-scan was performed, revealing a grade III AAST spleen laceration involving the inferior pole, associated with a subcapsular hematoma and several small intraparenchymal hematomas, with no active extravasation of contrast (fig. 1). No other injuries, namely bone fractures in the thoracic cage or involving abdominal solid organs, were observed. In light of the standard treatment of isolated splenic injuries in a stable pediatric patient, he remained in the hospital for clinical surveillance. Although clinically asymptomatic during hospital stay, Doppler ultrasound (US) performed 11 days after trauma demonstrated an increase of the subcapsular hematoma, with internal vascularization and feeding vessels in its vicinity suggesting PA formation (fig. 2). This finding was confirmed with computer tomography angiography (CTA) (fig. 3), which demonstrated an hypervascular structure within the cleft of the splenic laceration. The patient underwent angiography via right femoral artery approach. Selective angiography of the splenic artery with Simmons 5Fr catheter (Tempo, Cordis®, Miami, FL, USA) demonstrated the presence of a PA in the lower pole (fig. 4a). There was no evidence of other PAs or contrast extravasation within the splenic vascular bed. After superselective catheterization with 2.7Fr microcatheter (Progreat, Terumo®, Tokyo, Japan), transcatheter embolization was first performed with N-butyl-cyanoacrylate (NBCA, Glubran Tiss, GEM, Viareggio, Italy) prepared with lipiodol (Guerbert, Aulnay-sous-Bois, France)



Figure 1 – Contrast-enhanced coronal (a) and axial (b) CT. Splenic parenchyma laceration evolving the lower pole with subacute hematoma (arrow).

1/3 ratio. It was not possible to achieve a distal position of the microcatheter tip inside the PA so NBCA was chosen to allow a distal penetration of the embolic from a proximal injection. Afterwards, Onyx 18® (Covidien Inc., Plymouth, MN, USA) was used for a more proximal embolization (fig. 4b). Post-embolization angiography showed exclusion of the PA (fig. 4c). Follow-up US-Doppler examination performed on post-procedural day 5 documented the thrombosed PA, with no vascular abnormalities associated. (fig. 5). The patient underwent an uneventful recovery and was discharged 6 days after angioembolization.

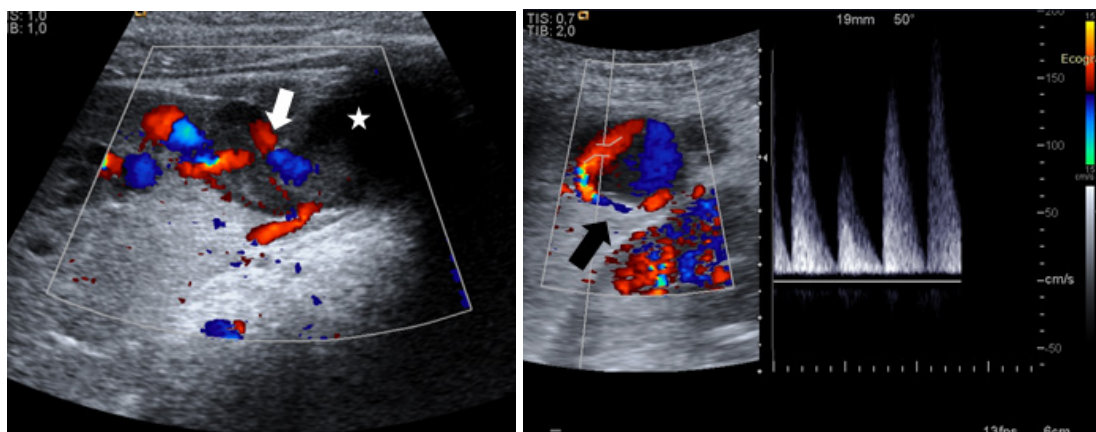


Figure 2 – Doppler-US. Subcapsular splenic hematoma (star) and areas of arterial flow in its vicinity (arrow, left) with yin-yang sign (arrow, right), suggesting pseudoaneurysm.

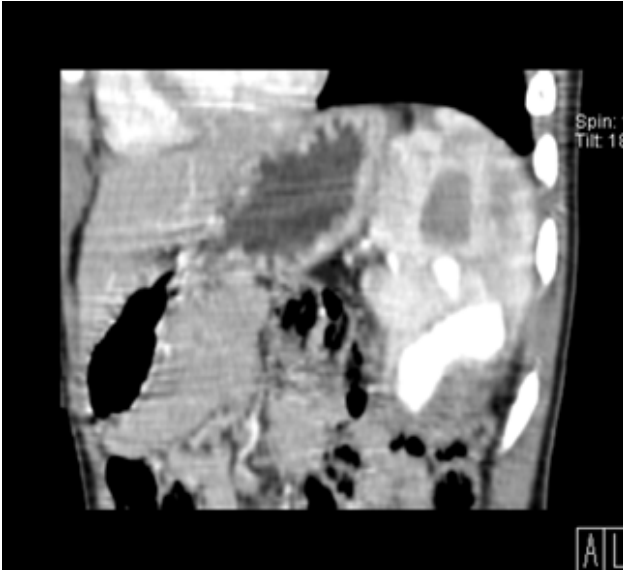


Figure 3 – Contrast-enhanced sagittal CT, arterial phase. Contrast extravasation within a contained sac confirming a large pseudoaneurysm (arrow) in the inferior splenic pole.

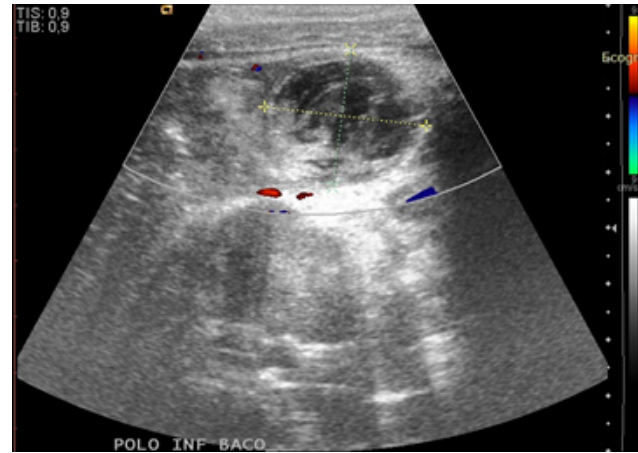


Figure 5 – Doppler US. Thrombosed pseudoaneurysm with no internal vascularization.

Case 2

A 12 year-old female with no significant past medical history presented herself to the referring hospital after sustaining an 8 meter fall, with blunt upper abdominal and facial trauma. The child was hemodynamically stable, and her vital signs were within normal limits on arrival to the emergency room, with a Glasgow Coma Scale (GCS) of 13. She presented several facial excoriations, a peri-orbital hematoma, lower lip laceration and upper superior abdominal tenderness upon physical exam. Her blood and urine tests showed elevated liver enzymes (AST – 807 u/L; ALT – 684 u/L) and microscopic hematuria. Her hemoglobin level was 7,6 g/dL requiring red blood cell transfusion. Abdominal CT scan showed moderate volume hemoperitoneum, grade III AAST liver laceration in the right lobe with no evidence of active extravasation of contrast and grade I AAST contusion of the right kidney. There were also several maxillofacial fractures demonstrated in head CT, treated during hospital stay. Non operative management was decided for the abdominal injuries. 11 days after trauma follow-up abdominal CT was performed, showing a 15mm arterial PA in the right hepatic lobe and early opacification of a portal branch in its vicinity, suggesting an arterio-portal fistula. The patient was referred to our institution for angiographic embolization. After right common femoral artery retrograde puncture, a 5Fr sheath-introducer catheter was placed. Careful observation of the CTA prior to the procedure revealed replaced origin of the hepatic artery directly from the abdominal aorta. Selective catheterization and subsequent angiography of the right hepatic artery were performed using a Simmons 5 Fr catheter (Tempo, Cordis®, Miami, FL, USA) confirmed the PA and the arterio-portal fistula arising from a segmental branch (fig. 6a). Superselective angiography of the left hepatic artery, also branching from the aorta, excluded vascular abnormalities (fig. 6b). A 2.7 Fr microcatheter (Progreat, Terumo®, Tokyo, Japan) was advanced coaxially through the Simmons catheter with superselectivation of the segmental branch feeding the PA and arterio-portal fistula. Due to the high-flow fistulous connection of the PA to the portal vein, embolization was performed with Onyx 18® (fig. 6c), and repeated contrast injection into the right hepatic artery demonstrated no filling of the pseudoaneurysm and exclusion of the arterio-portal fistula (fig. 6d). No major nor minor complications were registered after procedure. The child was transferred back to her referring hospital and was maintained under conservative

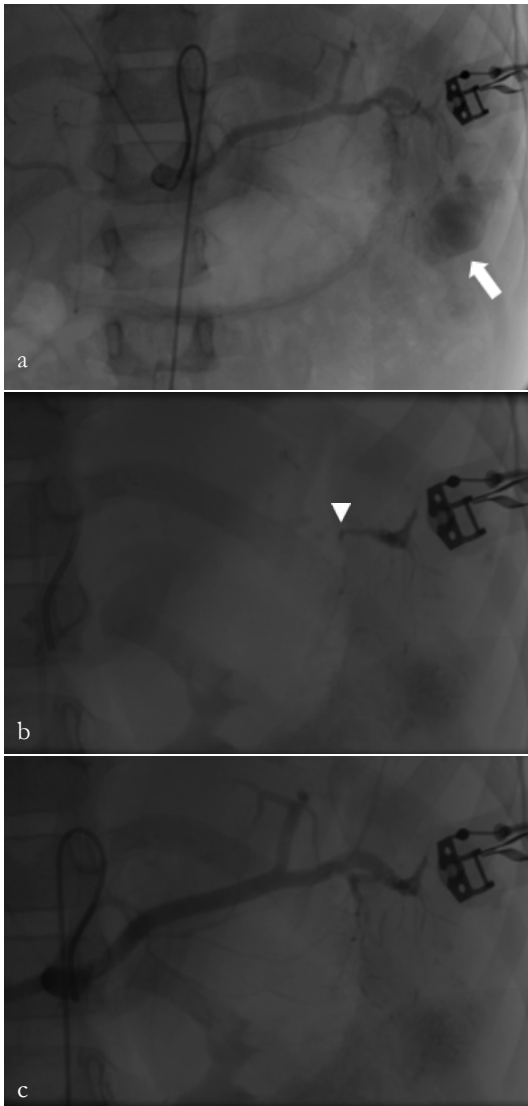


Figure 4 – a) Celiac arteriogram revealing the pseudoaneurysm (arrow) in the inferior splenic pole. b) Embolization with Onyx 18® (arrowhead). c) Celiac aortogram post-embolization demonstrating vascular exclusion of the pseudoaneurysm.

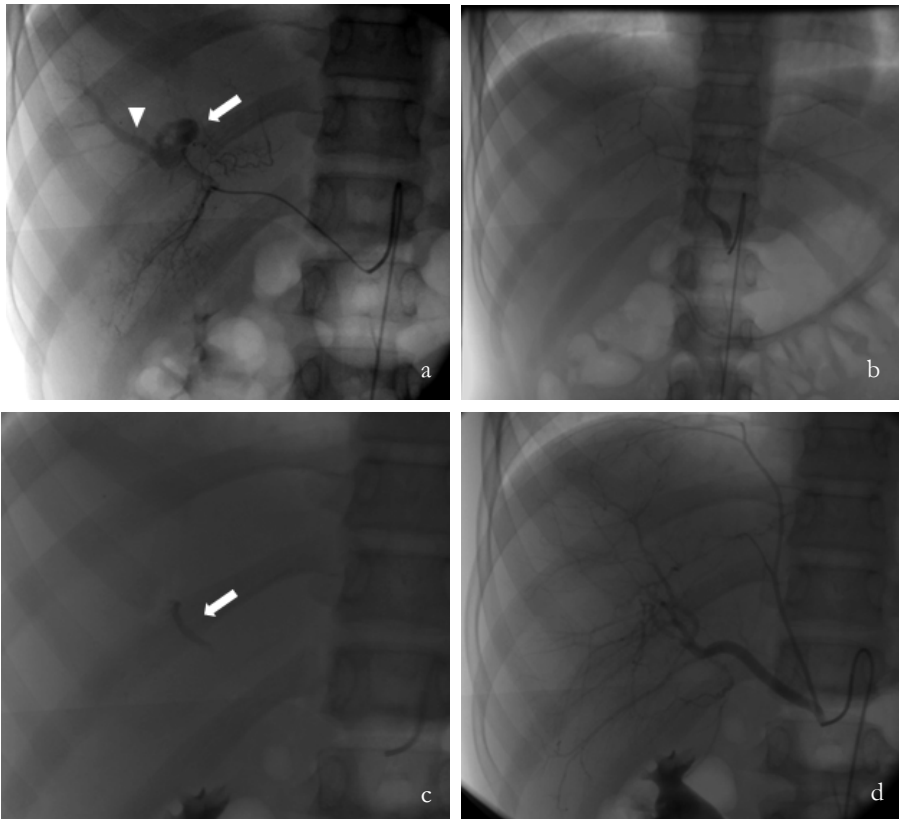


Figure 6 – a) Selective angiography from the right hepatic artery. Large pseudoaneurysm (arrow) arising from a segmental branch, associated with early opacification of a portal venous branch suggesting arteriovenous fistula (arrowhead). b) Selective angiography from the left hepatic artery. c) Embolization with Onyx 18® (arrow). d) Post-embolization angiogram from a common arterial trunk (right hepatic and phrenic arteries) with no opacification of the pseudoaneurysm or arteriovenous fistula.

treatment. Her liver enzymes returned to normal values (AST – 18 u/L; ALT – 39 u/L) and Doppler-US performed 2 days after endovascular intervention showed resolution of the vascular lesions. After an uneventful recovery, she was discharged 6 days after angioembolization. Follow-up CT performed 5 months later demonstrated vascular exclusion of the segmental branch with no evidence of PA or arterioportal fistula, with a patent portal vein.

Case 3

A 17-year old male adolescent was admitted to a regional hospital after sustaining a blunt lumbar trauma in a bicycle crash. Abdominal computed tomography was performed, revealing a grade III AAST injury in the right kidney, with parenchymal laceration and a large peri-renal hematoma (8,3x8cm) with no parenchymal contrast uptake in the upper third of the kidney. There was no evidence of collecting system lesion or active extravasation of iodine contrast. No other traumatic lesions were found in the abdominopelvic cavity. He was transferred to our institution for further management. On admission, he was conscious, hemodynamically stable, referring pain in the lumbar region. The patient's physical examination revealed only excoriations in the right lumbar region. His hemoglobin level was 12,8g/dl. The patient underwent non-operative management and proceeded to an uneventful clinical recovery. Ultrasound performed at day 7 after trauma revealed slight decrease in the size of the hematoma and no active bleeding (fig. 7). He was discharged home the day after. At day 15 after trauma, the patient was readmitted to his hospital with acute and intense right lumbar pain. His CBC showed a decrease in hemoglobin level (11,8g/dL). He underwent CTA imaging (fig.8), revealing a pericentimetric PA in the upper third of the

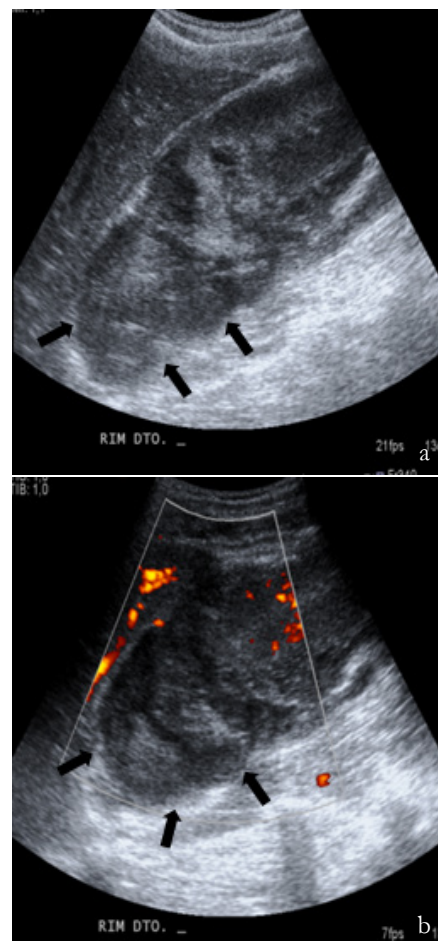


Figure 7 a) and b) - Doppler US. Peri-renal hematoma (arrows) with no vascular flow in the upper third of right kidney.

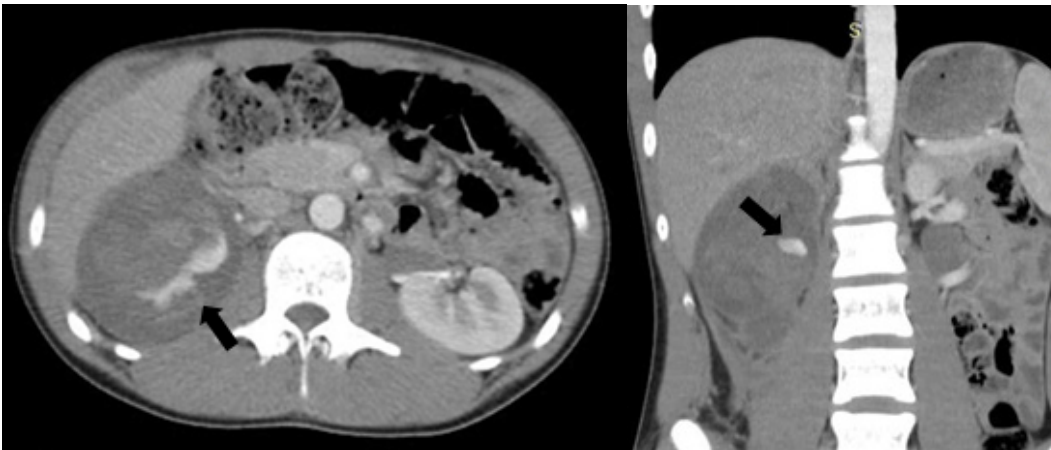


Figure 8 – Contrast-enhanced axial and coronal images. Contrast-filled sac with relation with a segmental branch of the right renal artery suggesting pseudoaneurysm (arrow)

right kidney with active extravasation of contrast. There was also a slight increase in the size of the peri-renal hematoma. He was readmitted to our department for angiography. Using a right femoral approach, a selective angiography of the right renal artery was performed, that demonstrated the PA in the posterior segmental artery with active extravasation of contrast, as well as 2 other millimetric PAs (fig. 9a). Due to the anatomic location, aneurysm size and poor stability of the microcatheter tip inside the aneurysm sac, detachable coils were selected for embolization. Two 0.018 inch detachable microcoils of 15mm in diameter were deployed through a 2.7Fr coaxial microcatheter (Progreat®, Terumo, Tokyo, Japan) after superselective catheterization of the larger PA. It was decided to manage conservatively the other PAs due to their small size and location. Control angiography revealed successful embolization of the pseudoaneurysm preserving vascular flow to the anteroinferior and inferior segmental arteries (fig.9b). CTA was performed 72 hours later, confirming the exclusion of the PA and preserved

vascular flow to the remaining segmental arteries, as well as thrombosis of the smaller PAs (fig.10). The patient was discharged 12 days after embolization. During follow-up, he developed arterial hypertension, controlled with an ACE inhibitor. MR imaging performed 14 months after treatment showed a small remaining right kidney with normal contrast uptake (fig.11). Further evaluation with dimercaptosuccinic acid (DMSA) renal scan showed decreased renal function in the right kidney (right kidney - 13%; left kidney – 87%). Although this could have been partially due to segmental angioembolization, we believe that the initial vascular injury to the upper hemikidney evidenced in post-trauma imaging studies played a major role in this clinical outcome. Selective angiography before embolization (fig. 9a) showed no parenchymal perfusion to the upper 2/3 of the right kidney, proving that vascular occlusion with devascularization of 2/3 of the affected kidney was already present before embolization.



Figure 9 – Selective angiograms from the right renal artery: a) prior to embolization, revealing a pseudoaneurysm arising from the posterior segmental artery (arrow), as well as smaller pseudoaneurysms arising from the inferior segmental artery (arrowheads); b) Post-embolization control reveals correct microcoil deployment with vascular exclusion of the pseudoaneurysm (circle).

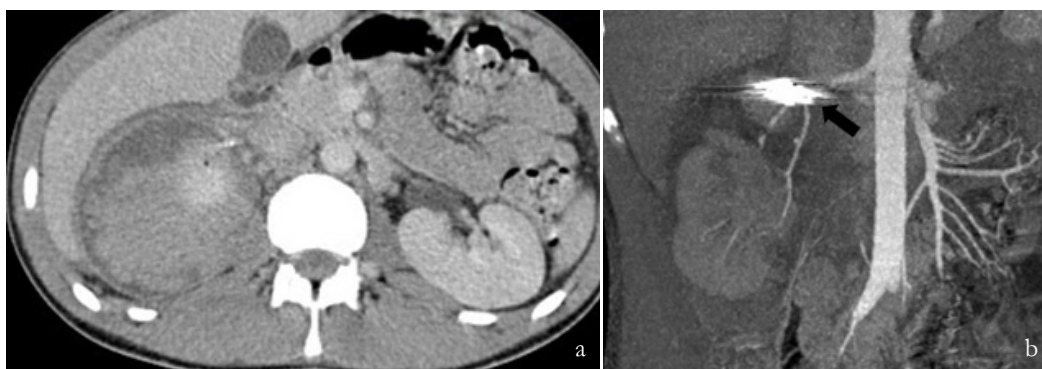


Figure 10 – Contrast-enhanced axial (a) and coronal (b) CT images confirming the exclusion of the PA with microcoils (arrow) and preserved vascular flow to the remaining segmental arteries.

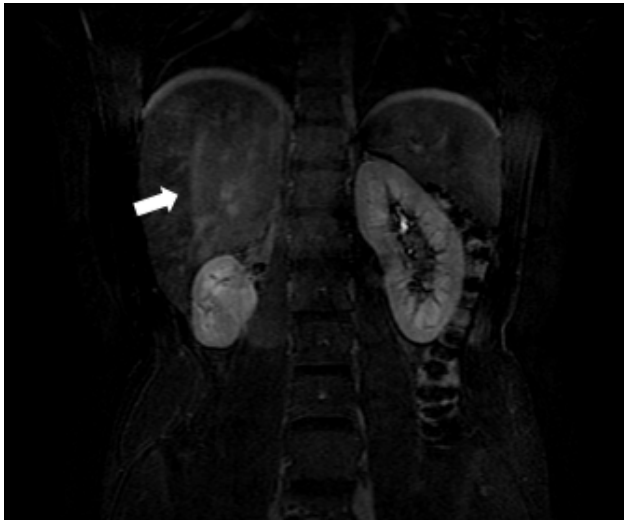


Figure 11 – T1-weighted contrast enhanced coronal MR image. Small remnant right kidney with normal contrast perfusion on venous phase.

Discussion

Abdominal trauma remains one of the leading causes of mortality and morbidity in the pediatric population. During the past decades, there has been a major shift from surgical to selective non operative management (NOM) of traumatic blunt abdominal injuries, and in hemodynamically stable children with blunt solid organ injury grade I to IV (American Association for the Surgery of Trauma organ injury scale, table 1), NOM of blunt abdominal solid organ injuries is currently the standard of care. Operative intervention, however, is still performed in patients with hemodynamic instability and/or active hemorrhage, often leading to removal of the injured organ with significant clinical consequences, such as a lifelong risk of overwhelming post-splenectomy infection, and potential progression to chronic renal disease after unilateral nephrectomy due to long-term vicarious glomerular hyperfiltration in the contralateral kidney.

Table 1 - The American Association for the Surgery of Trauma grading system for blunt injury of the liver, spleen and kidney

AAST blunt solid organ injury grade	Liver	Spleen	Kidney
I	Subcapsular haematoma: <10% surface area Laceration: <1 cm depth	Subcapsular haematoma: <10% of surface area Laceration <1 cm depth	Contusion Subcapsular haematoma: non expanding
II	Subcapsular haematoma: 10-50% surface area Intraparenchymal haematoma: <10 cm diameter Laceration: 1-3 cm parenchymal depth, <10 cm length	Subcapsular haematoma: 10-50% of surface area Intraparenchymal haematoma: <5 cm in diameter Laceration 1-3 cm in depth not involving trabecular vessels	Perirenal haematoma: non-expanding, confined to retroperitoneum Laceration: <1 cm depth, not involving the collecting system
III	Subcapsular haematoma: >50% surface area of ruptured subcapsular or parenchymal haematoma Intraparenchymal haematoma: >10 cm or expanding Laceration: capsular tear >3 cm parenchymal depth	Subcapsular haematoma: >50% of surface area or expanding Intraparenchymal haematoma >5 cm or expanding Laceration: >3 cm in depth or involving trabecular vessels	Laceration: >1 cm without extension into the renal pelvis or collecting system
IV	Laceration: parenchymal disruption involving 25-75% hepatic lobe or involves 1-3 Couinaud segments	Ruptured subcapsular or parenchymal haematoma Laceration: involving segmental or hilar vessels with major devascularisation (>25% of spleen)	Laceration: extending to renal pelvis or urinary extravasation Vascular: injury to main renal artery or vein with contained haemorrhage Segmental infarctions without associated lacerations
V	Laceration: parenchymal disruption involving >75% of hepatic lobe or involves >3 Couinaud segments (within one lobe) Vascular: juxtahepatic venous injuries (retrohepatic vena cava / central major hepatic veins)	Shattered spleen Hilar vascular injury with splenic devascularisation	Avulsion of renal hilum: devascularisation of a kidney due to hilar injury Ureteropelvic avulsions
VI	Vascular - hepatic avulsion		Complete laceration or thrombus of the main renal artery or vein

In adults sustaining blunt abdominal trauma injuries, angiographic selective embolization of solid organ vascular territories has increasingly been used as an adjunct to NOM allowing for control of hemorrhage and organ salvage with high success and safety rates reported by various authors, resulting in a decrease the NOM failure and allowing for an expansion of NOM indications to more severe trauma patients, such as blunt solid organ high grade injuries (blunt solid organ injury grade V), older age and associated injuries.^{4-6,10}

Splanchnic PAs in blunt abdominal trauma setting develop following a disruption in arterial wall continuity. Due to sustained arterial pressure, blood dissects into the tissues around the damaged vessel and forms a perfused sac contained only by the outer vascular layers or simply by soft-tissue structures around the vessel.¹ Its most common sites include the spleen, liver and kidney vascular beds. Although the majority of them are asymptomatic, they can present with abdominal pain, obstructive jaundice, hemobilia or hematuria.¹¹ PAs have been recognized as a significant risk factor for NOM failure due to delayed hemorrhage, a low incidence (5–25% of splenic, 0–3.9% of hepatic and 0–9% of renal injuries, respectively) but life-threatening complication of NOM,¹² manifesting as intra-abdominal or retroperitoneal bleeding and requiring emergency treatment. Regarding blunt splenic trauma, delayed rupture is seen in 5% to 6% of cases and is often attributed to the rupture of a posttraumatic splenic PA.^{13,14} A study by Schurr et al demonstrated splenic PA in 67% of adults who failed NOM.^{15,16} This relationship between failed NOM and posttraumatic PAs has led to the practice in several trauma centers of semi-elective angiographic embolization of splanchnic PAs if detected in imaging studies.

The significance of post-traumatic splanchnic PAs in the pediatric population is less well defined, as it has until recently not been emphasized as a significant complication following abdominal trauma. There is also a lack of understanding regarding the natural history of traumatic abdominal PAs which are often asymptomatic and may not develop until days or even months after the initial injury, rendering its detection very difficult. The overall incidence of splenic and hepatic PAs reported in previous series is 2-27%² and 1,7-25%,^{17,18} respectively. This discrepancy in numbers may be related to different strategies between pediatric trauma centers for follow-up imaging in children in NOM. However, there is a general consensus regarding the relationship between the frequency of PAs and the severity of blunt solid organ injury. For instance, in one series reporting 5,4% of splenic PAs and 1,7% of hepatic PAs in 101 children sustaining abdominal trauma, the incidence was significantly higher in grade IV injuries (17 and 27%, respectively).¹¹

Currently, there are no high-grade recommendations for the screening and management of abdominal PAs in children, and the role of embolization as an adjunctive procedure is still unclear, partially due to higher success rates for NOM of spleen, liver, and renal injuries. In a recent guideline consensus published in 2017 by the World Society of Emergency Surgery regarding splenic trauma,¹⁸ a level 2C evidence recommendation was attributed for angioembolization of post-traumatic splenic pseudo-aneurysms prior to patient discharge. PAs in these abdominal organs are thought to frequently self-tamponade and undergo spontaneous thrombosis due to children's more elastic parenchyma and a thicker capsule, especially with small size PAs. It is however

mentioned that some authors recommend follow-up imaging to be performed prior to discharge, and in case of persistence of splenic PAs, angioembolization should be performed. The fear of delayed splenic rupture and other late complications of PAs such as arteriovenous fistula formation were cited as the reason for embolization in hemodynamically stable children. This is also the case in adolescents of more than 13-15 years old, that should be managed according to adults' protocols (level 1C recommendation). In children of less than 13 years old that are more vulnerable to overwhelming post-splenectomy infection, angioembolization should be considered as well.¹⁸ Regarding other sites of PA formation, namely liver and kidney, only case reports have been described in the literature.¹⁹⁻²³ Most trauma centers opt to manage these cases with prophylactic angiographic embolization due to the risk of catastrophic bleeding, especially in larger PAs that don't resolve spontaneously or that increase in size in imaging follow-up, as in the first case presented in this paper, as the technique and success of angiographic embolization are quite satisfactory.

Overall, there is a lower risk of complications in the endovascular treatment of visceral PAs compared to surgical management, and mortality and major complications are rarely reported.^{12,19} Potential complications following selective angioembolization in children are similar to those in adults, namely arterial puncture site hematoma, contrast nephropathy catheter or guidewire-related arterial injury, target organ ischemia, non-target organ embolization, and intra-procedural rupture of the PA. Post-embolization syndrome is frequently reported, consisting of abdominal pain, nausea, ileus and fever, but it is usually self-limited and tends to resolve spontaneously in 6 to 9 days. Splenic infarction areas are frequent, and several quantitative studies²⁴⁻²⁶ have demonstrated preservation of the reticulo-endothelial function of the spleen after selective angioembolization as the trabecular distribution of the intraparenchymal splenic vessels allows for targeted embolization while preserving blood flow to noninjured areas of the organ. Regarding the liver vasculature, ischemia is a rare complication due to the dual blood supply from the hepatic artery and the portal vein.¹⁹ Delayed failure of embolization, although very rare, has been reported due to recanalization of the embolized vessel and reconstitution of arterial flow to the PA.¹

We presented 3 cases of abdominal PAs that didn't resolve with spontaneous thrombosis or demonstrated progression in size or signs of active bleeding after conservative management. In this clinical scenario, as the risk of life-threatening intra-peritoneal hemorrhage is significantly higher, we believe that the benefits of prophylactic embolization clearly outweigh its potential complications, as early embolization under controlled circumstances is obviously preferable to radiologic or surgical intervention to treat PA rupture with catastrophic bleeding.

Conclusions

NOM is now the mainstay of treatment for clinically stable children with solid organ injury grade I-IV (AAST) after blunt abdominal trauma, with a success rate over 90% of cases. Delayed catastrophic bleeding requiring emergency treatment is, however, a potential life-threatening complication of this management approach, usually attributed to rupture of a splanchnic post-traumatic pseudoaneurysm. The capability of selective angioembolization to decrease NOM failure rate

in adults with blunt abdominal solid organ injuries has been demonstrated. Although currently no high-level evidence-based guidelines exist regarding the management of abdominal PAs in children, we believe that selective angioembolization should be strongly considered as an adjunctive treatment to NOM in persistent or evolving posttraumatic abdominal PAs, as demonstrated in the cases presented in this article.

Received / Recebido 03/12/2017

Acceptance / Aceite 04/02/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.
Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Saad NEA, Saad WE, Davies MG, et al. Pseudoaneurysms and the role of minimally invasive techniques in their management. *Radiographics*. 2005;25:173-90.
2. Martin K, Vanhouwelingen L, Bütter A. The significance of pseudoaneurysms in the nonoperative management of pediatric blunt splenic trauma. *J Pediatr Surg*. 2011;46:933-7.
3. Iacobellis ABF, Villamaina URE. Non operative management of blunt splenic trauma: a prospective evaluation of a standardized treatment protocol. *Eur J Trauma Emerg Surg*. 2016;42:1-6.
4. Haan JM, Bochicchio G V, Kramer N. Nonoperative management of blunt splenic injury: a 5-year experience. *J Trauma*. 2005;58:492-8.
5. Cheynel N, Loffroy R, Guiu B, et al. Transcatheter arterial embolization of splenic artery aneurysms and pseudoaneurysms: short- and long-term results. *Ann Vasc Surg*. 2008;22:618-26.
6. Finley DS, Hinojosa MW, Paya M, et al. Hepatic artery pseudoaneurysm: a report of seven cases and a review of the literature. *Surg Today*. 2005;35:543-7.
7. Lynn KN, Werder GM, Callaghan RM, et al. Pediatric blunt splenic trauma: a comprehensive review. *Pediatr Radiol*. 2009;39:904-16.
8. Soudack M, Epelman M, Gaitini D. Spontaneous thrombosis of hepatic posttraumatic pseudoaneurysms: sonographic and computed tomographic features. *J Ultrasound Med*. 2003;22:99-103.
9. Khoo I, Lim F, Miao P, et al. Prophylactic embolization of hepatic artery pseudoaneurysm after blunt abdominal trauma in a child. *J Pediatr Surg*. 2010;45:837-9.
10. Bhullar IS, Frykberg ER, Siragusa D, et al. Selective angiographic embolization of blunt splenic traumatic injuries in adults decreases failure rate of nonoperative management. *J Trauma Acute Care Surg*. 2012;72:1127-34.

Current available data in the literature have also reported very satisfactory results regarding this matter, but are based only on cohort studies and case reports. Prospective randomized-controlled trials are lacking in order to define its incidence, natural history and management, as well as which patients will benefit from earlier angioembolization.

11. Durkin N, Deganello A, Sellars ME, et al. Post-traumatic liver and splenic pseudoaneurysms in children: diagnosis, management, and follow-up screening using contrast enhanced ultrasound (CEUS). *J Pediatr Surg*. 2016;51:289-92.
12. Schuster T, Leissner G. Selective angioembolization in blunt solid organ injury in children and adolescents: review of recent literature and own experiences. *Eur J Pediatr Surg*. 2013;23:454-63.
13. Haan JM, Scalea TM. Blunt splenic injuries in the adolescent trauma population: the role of angiography and embolization. *JEM*. 2011;41:21-8.
14. Skattum J, Gaarder C, Aksel P. Splenic artery embolisation in children and adolescents - an 8 year experience. *Injury*. 2014;45:160-3.
15. Schurr M, Fabian T, Gavant M, et al. Management of blunt splenic trauma: computed tomographic contrast blush predicts failure of nonoperative management. *J Trauma*. 1995;39:507-13.
16. Kittaka H, Yagi Y, Zushi R, Hazui H, Akimoto H. The investigation of posttraumatic pseudoaneurysms in patients treated with nonoperative management for blunt abdominal solid organ injuries. *PLoS One*. 2015;10:1-12.
17. Safavi A, Beaudry P, Jamieson D, Murphy JJ. Traumatic pseudoaneurysms of the liver and spleen in children: is routine screening warranted? *J Pediatr Surg*. 2011;46:938-41.
18. Coccolini F, Montori G, Catena F, Kluger Y, Biffi W, Moore EE, et al. Splenic trauma: WSES classification and guidelines for adult and pediatric patients. *World J Emerg Surg*. 2017;12:1-26.
19. Kiankhooy A, Sartorelli KH, Vane DW, Bhavne AD. Angiographic embolization is safe and effective therapy for blunt abdominal solid organ injury in children. *J Trauma*. 2010;68:526-31.
20. Yamaçake KGR, Lucon M, Lucon AM, et al. Renal artery pseudoaneurysm after blunt renal trauma: report on three cases and review of the literature. *Sao Paulo Med J*. 2013;131:356-362.
21. Garg A, Gokhale A, Garg P, et al. Endovascular treatment of a delayed renal artery pseudoaneurysm following blunt abdominal trauma. *Urol J*. 2007;4:184-6.
22. Hardcastle TC, Reitz D, Hollander Dd, Rodseth R, Muckart DJ. Posttraumatic intrahepatic pseudoaneurysm in a child managed by coil angioembolization: a case report and literature review. *J Pediatr Surg*. 2010; 45:1-3.
23. Ong C, et al. Primary hepatic artery embolization in pediatric blunt trauma. *J Pediatr Surg*. 2012;47:2316-20.
24. Skattum J, Jeanette R, Loekke V, Larsen T, Grete A, Aaberge IS, et al. Preserved function after angioembolisation of splenic injury in children and adolescents: a case control study. *Injury*. 2014;45:156-9.
25. Sclafani S, Shaftan G, Scalea T. Nonoperative salvage of computed tomography-diagnosed splenic injuries: utilization of angiography for triage and embolization for hemostasis. *J Trauma*. 1995;39:818-27.
26. Schimmer JAG, Steeg AFW Van Der, Zuidema WP. Splenic function after angioembolization for splenic trauma in children and adults: A systematic review. *Injury*. 2016;47:525-30.
27. Tinkoff G, Esposito TJ, Reed J, Kilgo P, Fildes J, Pasquale M, Meredith JW. American association for the surgery of trauma organ injury scale I: spleen, liver, and kidney, validation based on the national trauma data bank. *Journal of the American College of Surgeons*. 2008;207:646-55.

The Role of Radiology in Detecting Prosthetic Breast Implant-Related Complications

O Papel da Radiologia na Monitorização das Complicações Relacionadas com as Próteses Mamárias

Willian Schmitt, João Morais Coelho, João Lopes, José Carlos Marques

Serviço de Radiologia, Hospital Prof. Doutor
Fernando Fonseca, Amadora, Portugal

Address

Willian Schmitt
Rua Manuel Moreira de Barros
618B R305
4400-346 Vila Nova de Gaia, Portugal
e.mail: schmitt.wr@gmail.com

Abstract

Given the growing number of breast implants, both for aesthetic and reconstructive purposes, it is imperative for the Radiologist, to know the different types of surgical procedures involved, as well as the various types of implants available. Several types of early or late complications can result from this. The prosthesis rupture represents one of the most frequent kinds of late complication and constitutes the foremost cause of its removal. Since its clinical manifestation may be absent in up to 50% of cases, Radiology plays a central role in its monitoring. In Portugal, MR surveillance is not recommended by the national health program. Ultrasound examination along with screening mammography is the usual preferred method.

Thus, although breast implants imaging constitutes a low percentage of the day-to-day care activity of a Radiologist, the knowledge of the different imaging findings in the multimodality imaging used is crucial for early diagnosis of these complications and to provide the best patient care possible.

Keywords

Breast implants; Capsular rupture; Breast augmentation; Breast reconstruction.

Resumo

Atendendo ao número crescente de utilização de próteses mamárias, quer na cirurgia mamária estética, quer reconstrutiva, é fundamental que o Radiologista conheça os diferentes tipos de procedimentos cirúrgicos envolvidos, bem como os vários tipos de próteses disponíveis. A colocação de próteses mamárias não é um procedimento inócuo. Vários são os tipos de complicações descritas, podendo ocorrer precocemente, habitualmente no período pós-operatório ou num período mais tardio. A rutura da prótese representa um dos tipos de complicações tardias mais frequentes e constitui a principal causa da sua remoção. Uma vez que a manifestação clínica da rutura protésica pode estar ausente em até 50% dos casos, a Radiologia desempenha um papel central no seu diagnóstico. Em Portugal, a vigilância por ressonância magnética destas complicações não é recomendada pela Direção Geral de Saúde, sendo apenas recomendada a realização complementar de ecografia quando do estudo mamográfico de rastreio.

Assim, embora a avaliação de imagem das próteses constitua uma baixa percentagem da atividade diária de um Radiologista, o conhecimento dos diferentes achados de imagem nas diferentes técnicas utilizadas é crucial para o diagnóstico precoce destas complicações.

Palavras-chave

Próteses mamárias; Ruptura capsular; Mamoplastia; Reconstrução mamária.

1. Introduction

Initiated in the 19th century (Czérny),¹ breast-modifying surgery using synthetic material observed a significant progress in the 60s, with the development of silicone breast implants.² Whether for aesthetic or reconstructive reasons, the use of breast implants has been increasing. Currently, the breast enhancement mammoplasty is the aesthetical surgical procedure most commonly performed, with about 300,000 procedures performed in the US in 2016.³ The increase in the use of breast implants for breast reconstruction is directly related to the increase in the number of cases of immediate reconstruction, during the surgical procedure of the mastectomy.⁴ The radiologic evaluation of mammary implants constitutes a low percentage of daily care activity of a radiologist, so this

article aims to describe and illustrate the different types of complications and their translation in the various imaging techniques.

2. Surgical Procedure

The surgical technique, namely the type of incision and the plane of dissection, is individualized and performed according to the anatomy and the preferences of each patient, the experience of the surgeon and the type of surgery (aesthetic or reconstructive, primary or revision). In breast augmentation mammoplasty the different types of approach are: inframammary, periareolar, transaxillary or, rarely, transumbilical (Figure 1).

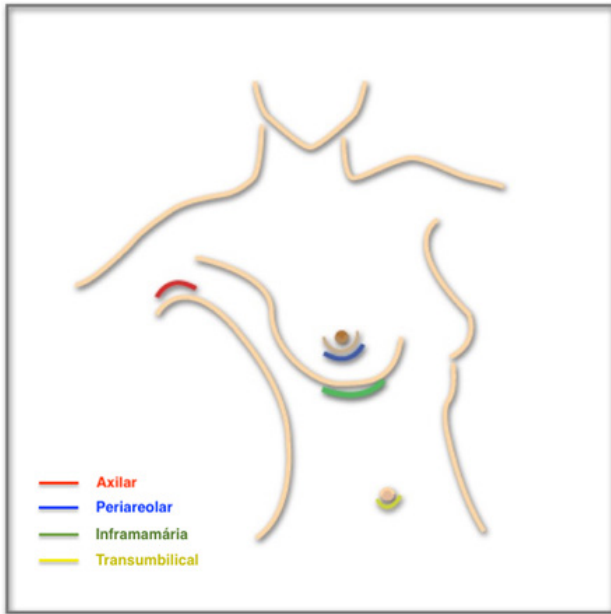


Figure 1 – Illustration of different types of incision.

The inframammary approach was the first to be described, allowing, like the inframammary and axillary incision, access to all dissection planes. This provides easy access, under direct visualization, facilitating the precise placement of the implant. However, given the surgical scar, new types of approaches have been developed. The periareolar approach followed, with the advantage of a potentially better scar from the aesthetic point of view. However, it only allows limited exposure of the surgical loca, so it is not advised in patients with small areolae as it may limit the placement of large volume implants and, due to the greater proximity to the ductal system, it is associated with an increased risk of infection. In the transaxillary approach the dissection can be performed either blindly or with the use of endoscopy. When performed blindly, it presents a greater risk of hematoma and nerve damage. The transumbilical access is used exclusively for the placement of saline implants. In this technique, the dissection of the prosthetic loca is technically demanding and may, as in the axillary approach, be performed by endoscopy or blindly.^{5,6}

Breast reconstruction is a commonly used surgical procedure after breast carcinoma mastectomy or prophylactic mastectomy in high-risk women.

The placement of implants is a reconstructive option that can be performed as an isolated technique (eg: skin sparing

mastectomy with immediate reconstruction with implants) or associated with other techniques such as:

- pedicled flaps (eg, myocutaneous flap of the *latissimus dorsi* muscle, myocutaneous flap of the *rectus abdominis* muscle - TRAM);
- free flaps (eg: deep inferior epigastric artery perforator flap - DIEP, free TRAM flap).

In breast reconstruction, the approaches vary according to the technique of tumor resection.

The technical description of these options goes beyond the objectives of this article, but its knowledge by the radiologist is essential for their correct interpretation.

Breast implants can be placed in several planes of dissection: retro-glandular, subfascial, retro-pectoral or in “dual-plane” (Figure 2).

The retro-glandular plane allows a less painful recovery, with greater ease of dissection. In order to achieve this, an amount of glandular tissue is required to cover the prosthesis in order to obtain better aesthetic results (Figure 3). In contrast, it is associated with a higher incidence of capsular contracture when compared to the other planes. The subfascial plane is a potential space between the pectoralis major and anterior serratus muscles and their corresponding fascia. This



Figure 3 – Pre and post surgery of breast augmentation mammoplasty with a retroglandular textured silicone implant placed by inframammary approach

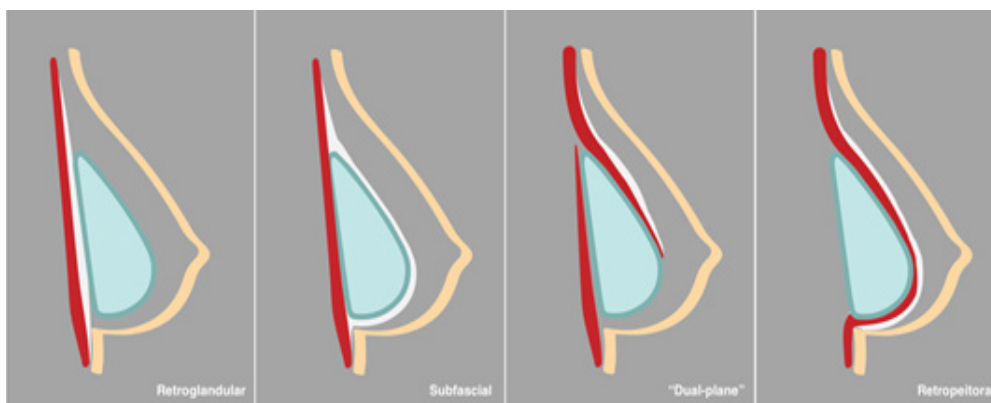


Figure 2 – Illustration of various dissection planes.

technique is similar to the retro-glandular one, granting, however, a greater coverage of the prosthesis. The retro-pectoral plane can be divided into totally submuscular, the least used considering the risk of incorrect positioning of the prosthesis, and the “dual-plane” in which the implant is covered by the pectoralis major muscle in the upper portion and by the mammary gland in the lower portion. This approach is usually used in cases where the subcutaneous tissue is scarce and in cases of capsular contracture revision mammoplasty, being associated with a more painful recovery. Compared with the retro-glandular approach there is a lower risk of capsular contracture.^{5,6}

3. Types of Implants

Various methods of breast modification have been described over time. The first reports describe the free injection of paraffin and, later, liquid silicone (Figure 4).

For some time, autologous transplant of adipose tissue has also been widely performed, however, given the associated risk of fat necrosis, the accuracy of lesion detection in the mammographic study was limited and so it was progressively abandoned.

In the 1960s, the first silicone implants were developed, which can be categorized into five generations. In parallel, saline implants were also developed, consisting of an inflatable envelope, which is filled with saline solution postoperatively.^{6,7} The knowledge of the different types of implants is fundamental in the interpretation of potential complications. The mammary implants can be classified according to their overall shape (anatomical or round), to the external surface (textured or smooth), the profile (high, medium or low), their content (saline or silicone) and the number of lumens (single or double).

The current silicone implants feature an external silicone elastomer shell filled with silicone gel. These can also be covered by a layer of polyurethane that is associated with a lower risk of contracture. Fifth generation prostheses

consist of a viscous silicone gel with high cohesiveness, being associated with a lower incidence of rupture.⁵ The most commonly used are those of single lumen, textured and filled with highly cohesive silicone gel.

4. Complications

Complications related to implants can be divided according to the time they take to be present.

4.1. Early complications

In the immediate postoperative period, complications related to the surgical procedure may lead to surgical reintervention. In this period, clinical evaluation plays a fundamental role, with imaging tests reserved to confirm clinical suspicion, to assess its extent and possibly to guide treatment. These complications include infection and hematoma.⁸

4.1.1 Infection

The rate of infection described after augmentation mammoplasty is 2-2.5% of the cases, slightly higher than in oncological surgery.⁹ The main related symptoms are pain, edema and erythema. Ultrasound may reveal the presence of an abscess, translated by the presence of a heterogeneous collection. In magnetic resonance imaging (MRI), findings such as skin thickening, interstitial edema, peripheral enhancement of the implant and peri-prosthetic collections suggest this diagnosis.

4.1.2 Hematoma

Hematoma formation often occurs in the immediate postoperative or post-trauma period. The mammographic study may show the presence of a well-defined hyperdense area, while the ultrasound study reveals the presence of a heterogeneous, multi-segmented collection (Figure 5).

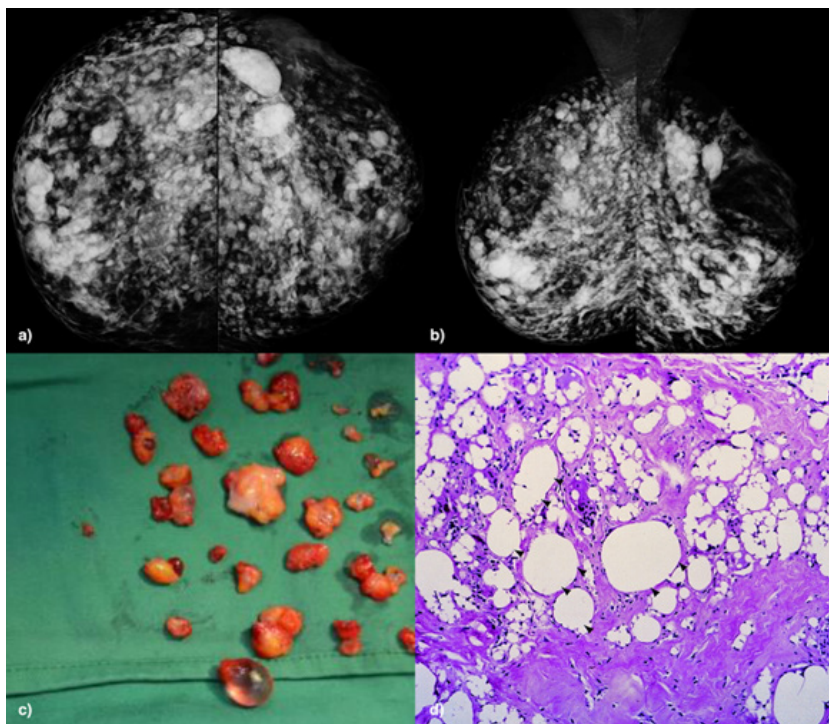


Figure 4 – A woman with a history of free silicone injection, showing the presence of multiple silicone granulomas (siliconomas), bilaterally, evidenced in the mammographic study (a and b), macroscopic (c) and histological (d) ones.

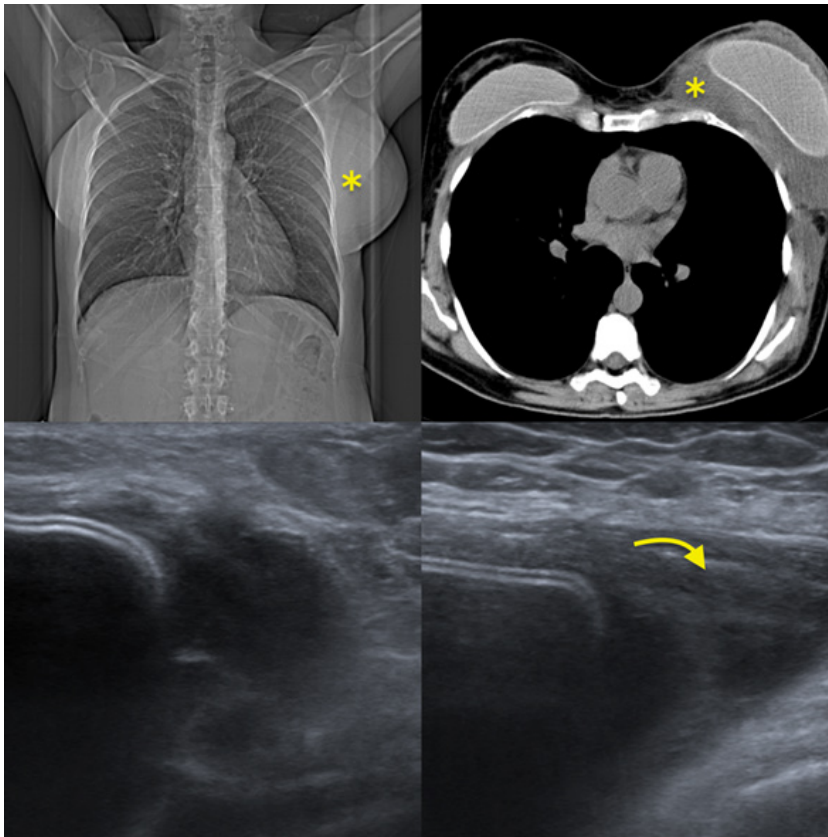


Figure 5 – A 46-year-old female submitted to augmentation mammoplasty, with markedly developed breast asymmetry during the postoperative period. The tomodensitometric study revealed the presence of voluminous periprosthetic collection (asterisk), translated in the ultrasound study by multiseptated collection suggestive of hematoma. An eco-guided drainage of this hematoma was performed, with a favorable evolution of the clinical picture.

4.2. Late complications

In the medium / long term, complications specifically related to breast implants arise.

4.2.1. Capsular contracture

After the placement of a breast implant, there is a foreign body reaction, with the formation of a fibrous capsule at its periphery. When this fibrotic response is excessive, capsular contracture occurs, one of the most frequent complications.⁸ This occurs more frequently in retro-glandular prostheses (8.6%), with a smooth surface.¹⁰ Its diagnosis is mostly clinical, with breast distortion which can be associated with local pain and inflammation. The ultrasound study may demonstrate the fibrous thickening of the capsule, whereas the mammographic study may show its morphological alteration, as well as the presence of peri-prosthetic calcifications. The latter are not a pathognomonic finding of capsular contracture, being very often related to the age of the prosthesis.^{8,11}

4.2.2. Capsular rupture

4.2.2.1 Mammography

It is the most widely used imaging method in mammary evaluation, mainly as a screening method, and as such, may be the first method to identify a possible complication. For screening purposes, additional incidences of Eklund should be performed in order to displace the prosthesis.¹² Some isolated reports of rupture of the prosthesis after compression are described, however they are thought to be related to previous intracapsular ruptures.¹³

When compared to other imaging methods, mammography has the lowest sensitivity for detecting ruptures (11-69%). This is mainly due to the fact that the prostheses are extremely radiodense, preventing the evaluation of their

internal content and, as such, the diagnosis of intracapsular rupture.

First, the location of the prostheses should be recognized, followed by analysis of their contours (Figures 6 and 7).

The appearance of undulations, focal herniations, morphological asymmetry or periprosthetic calcifications are unspecific findings but may be the first evidence of loss of prosthesis integrity. These should lead to further examinations to continue the investigation of eventual rupture.^{12,14}

Although little sensitive in the detection of intracapsular rupture, the mammography is very useful in the detecting extracapsular silicone, represented by the presence of a high density asymmetry in the parenchyma (Figures 8 and 9).

In the absence of rupture history or revision of the prosthesis, the presence of silicone outside the outer capsule means the presence of extracapsular rupture and, by extension, intracapsular rupture. In these cases it is not mandatory to perform other imaging methods for diagnostic confirmation, except for the purpose of investigating the contralateral prosthesis prior to surgical intervention.^{14,15}

Sometimes the extracapsular silicone can simulate larger lesions, forming granulomas, which may have spiculated contours. In these cases, a high level of suspicion is necessary in order to avoid unnecessary biopsies (Figure 10).

The extracapsular silicone may also extend along the fascia of the pectoralis major muscle or subcutaneous tissue to the axillary lymph nodes. However, it should be noted that the isolated presence of enlarged axillary lymph nodes with evidence of silicone inside is not pathognomonic of extracapsular rupture. By the so-called gel bleeding effect, small unpolymerized silicone molecules are able to transpose the outer capsule over time and are consequently drained by the lymphatic system. Thus, the isolated presence of enlarged axillary nodes with increased density, justify the

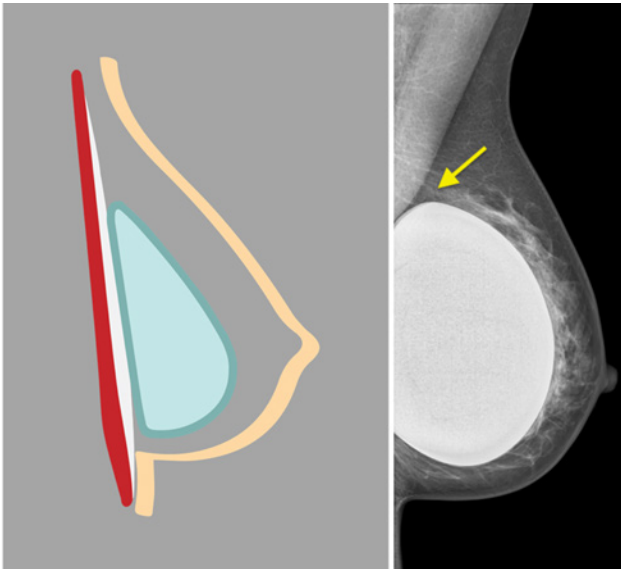


Figure 6 – Illustration and mammographic study on the mid-lateral-oblique incidence (MLO) demonstrating the presence of a silicone implant located in the subglandular plane, anterior to the pectoral muscle (arrow).

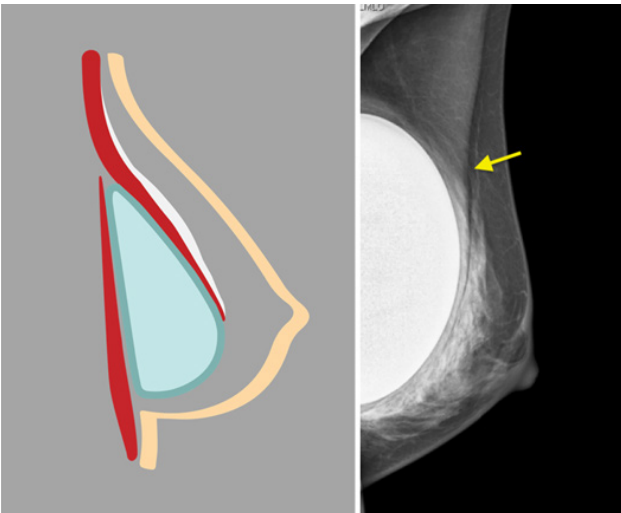


Figure 7 – Illustration and mammographic study on the MLO incidence of retromuscular silicone implant (arrow), in this case in “dual-plane”, only with partial dissection of the pectoralis major muscle.

complementary evaluation with ultrasound or MRI for the investigation of eventual rupture.¹⁴

4.2.2.2 Ultrasound

In Portugal, the ultrasound study is part of the mammary evaluation of women with breast implants, as mentioned in the DGS standard.¹⁶ Compared to mammography, it has a

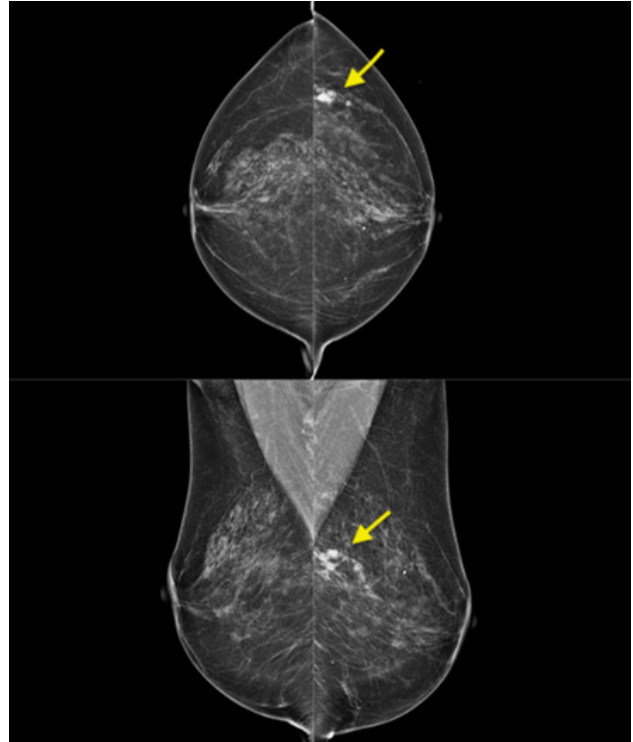


Figure 8 – A 75-year-old woman with a history of reduction mammoplasty with implant complications. The mammographic study revealed the presence of a high density asymmetry in the super-external quadrant of the left breast.

greater sensitivity in the detection of complications, being, however, inferior to MR (30-75%).^{14,17,18,19} This discrepancy can be explained by the fact that the ultrasound is operator-dependent and its execution varies with the technicians, general radiologists or radiologists differentiated in senology. The ultrasound study should be optimized for implant evaluation, with the selection of appropriate focus and depth. Its evaluation should be performed with a high frequency probe (7-15 Mhz), although a lower frequency may be useful in cases of larger implants.

The knowledge of the normal ultrasound aspect of the breast implants is fundamental in their evaluation. These are externally delimited by a regular trilaminar line, which progressively molds to the loca created during the surgical procedure, forming radiating folds, which should not be mistaken for signs of intracapsular rupture. This outer trilaminar contour is formed externally by a hyperechogenic line, which represents the outer face of the fibrous capsule, an innermost line representing the inner face of the elastomer shell and an intermediate line that translates the interface of these two components (Figure 11).

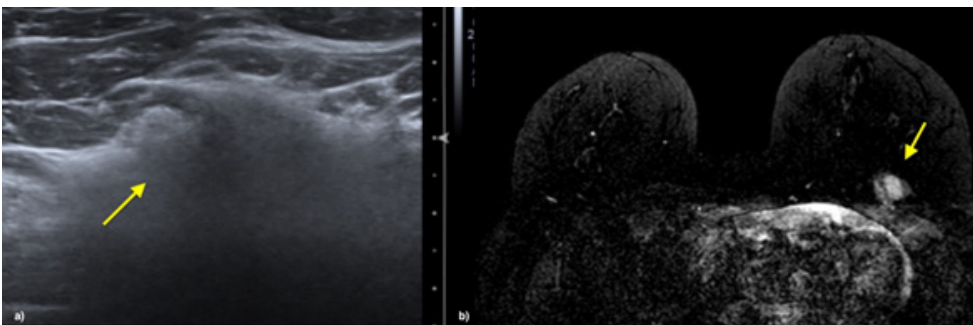


Figure 9 – A complementary ultrasound study (a), which demonstrated the presence of the “snowstorm” signal was performed, and the MRI (b) study revealed the presence of hypersignal in the selective sequence for the silicone. These findings are compatible with free silicone, these characteristics being described later in this article.

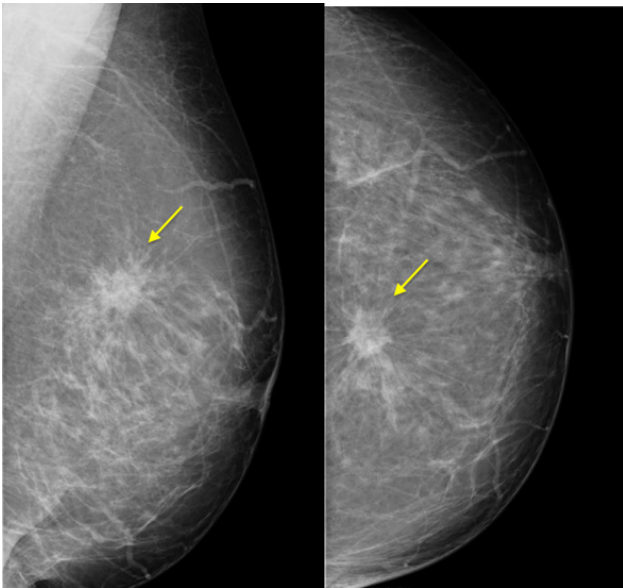


Figure 10 – Woman with a history of implants removal due to complication. The mammographic study revealed the presence of a suspicious lesion, with spiculated contours, and the biopsy result was negative. The MRI study revealed the presence of a non-capturing lesion, with a positive sign for silicone.



Figure 11 – Ultrasound study showing external trilaminar line of a silicone implant.

The demonstration of a regular trilaminar line during implant evaluation has, in the great majority of cases, a good correlation with its integrity.¹⁴

The ultrasound study of implants should include evaluation of their contours, their luminal content and the presence of free silicone or granulomas, both in the mammary parenchyma and axillary lymph nodes.¹⁵

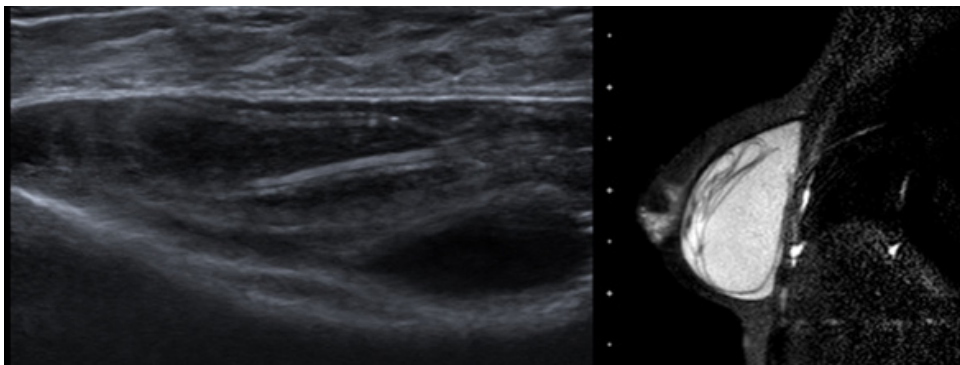


Figure 12 – A 40-year-old woman with a history of breast augmentation 12 years ago. The ultrasound study demonstrated the presence of several curvilinear lines inside the implant (ladder sign), in correspondence with the “linguini” sign in MRI, translating the presence of intracapsular rupture.

The most reliable sign of the integrity of the implant is the demonstration of an anechoic and homogeneous luminal content.¹³

The presence of several horizontal or curvilinear lines inside the implant, forming the stepladder sign, are the most reliable sign of intracapsular rupture, in which there is a greater collapse of the envelope, being equivalent to the sign of the “linguini” on MRI (Figure 12).⁸

Other signs such as the “lock sign” or the “subcapsular line” should also alert the radiologist to the presence of this type of rupture, representing earlier stages of the rupture. The lock sign occurs by expanding the apex of a radiating fold by extruding a small amount of silicone at this level. The sign from the subcapsular line represents a slightly posterior stage, where there is a greater accumulation of silicone in the space between the fibrous capsule and the inner membrane (Figure 13).

In the investigation of a possible rupture, potential pitfalls should have been taken into account. The presence of reverberation artifacts, with the presence of several echogenic lines parallel to the capsule-elastomer complex, should not be confused with the presence of rupture. This artifact is often conditioned by the excessive compression of the prosthesis, being eliminated with a softer compression.

The presence of a heterogeneous content may simulate the presence of rupture, especially in the 5th generation implants with silicon gel of high cohesiveness. Radial folds may also mimic this type of complication, however, since they represent only an invagination of the envelope, it is important to follow the path from the apex to the margin of the implant (Figure 14).

The presence of any signs suggesting intracapsular rupture should alert the radiologist to the eventual presence of associated extracapsular rupture. For this, the presence of free silicone in the mammary parenchyma, translated by the sign of the “snowstorm”, the most sensitive and specific signal of rupture on the ultrasound,¹⁴ should be investigated. This consists of a marked increase in echogenicity with loss of the parenchymal interface, conditioned by the dispersion of the ultrasonic beam caused by the silicone. The free silicon can still be absorbed by the lymphatic system, identifying the presence of this same sign in the axillary ganglia. As previously described, its presence is not diagnostic of rupture due to the existence of the gel-bleeding phenomenon, which is currently less frequent with the use of the latest generation silicone implants (Figure 15).

Depending on the amount of free silicone, it may form granulomas, the imaging spectrum of which can vary from an ultrasonographically simple cyst to a typical “snowstorm”

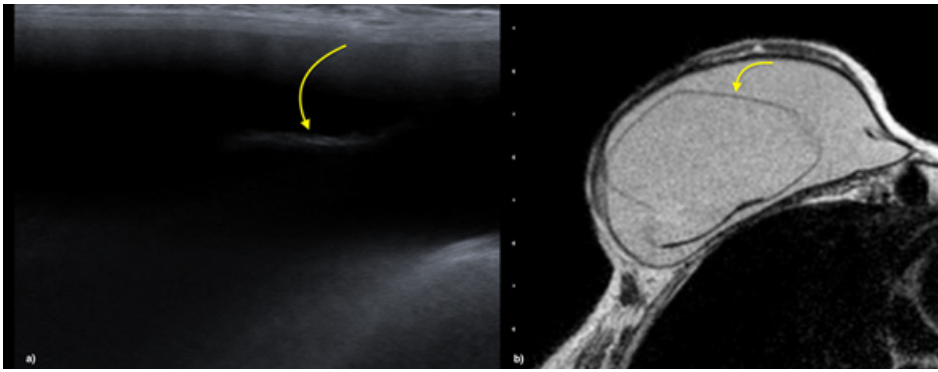


Figure 13 – An 85-year-old woman with a history of bilateral mastectomy for bilateral breast carcinoma and reconstruction with implants 20 years ago. In the ultrasound study (a) the loss of the usual trilaminar contour was verified, identifying the presence of the “subcapsular line” signal (arrow). The MRI study was consistent (b), revealing detachment of the internal membrane of the implant, being compatible with intracapsular rupture. This should not be mistaken with the presence of periprosthetic fluid, reason why it is crucial to evaluate the trilaminar line.

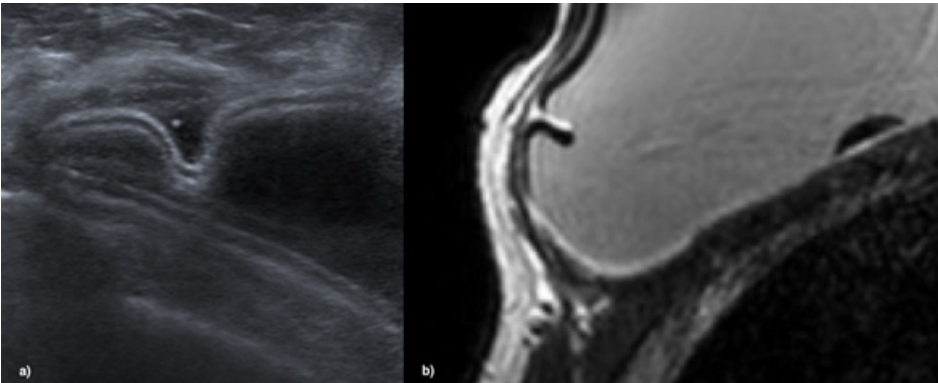


Figure 14 – Ultrasound (a) and MRI (b) showing the presence of a radiating fold.

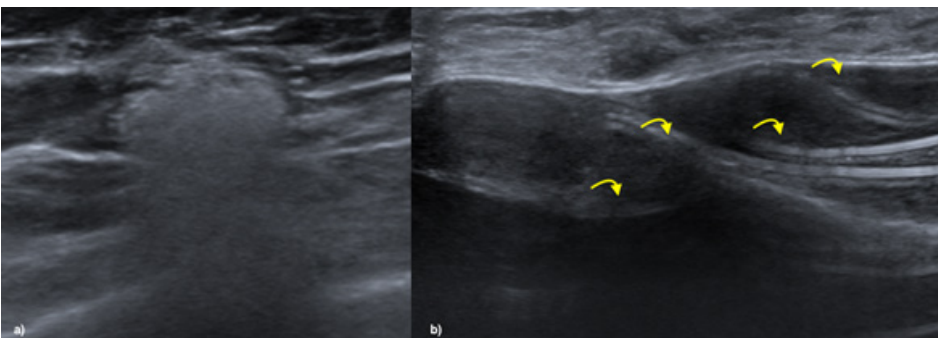


Figure 15 – A 40-year-old woman with a history of breast augmentation. The ultrasound study identified the presence of axillary ganglion with sign of the “snowstorm”, suggestive of extracapsular rupture, confirmed by the presence of the “ladder” sign in the homolateral implant, translating intracapsular rupture.

signal, or may present suspect ultrasound characteristics, resulting sometimes in a biopsy.

If the ultrasound findings are not categorical or if the evaluation has been impaired by the presence of granulomas related to previous ruptures, a complementary MRI study should be performed (Figure 16).

4.2.2.3 Magnetic Resonance

MRI is the most sensitive and specific imaging method for the detection of rupture, estimated at 72-84% and 85-100%, respectively.^{8,14}

This modality is used mainly in the evaluation of silicone implants, since the rupture of the saline implants in most cases is clinically evident and the fact that some expanders constitute a contraindication to its realization (Figure 17).

The study should be performed with a dedicated coil in an apparatus with a magnetic field of at least 1.5T, and its protocol should include selective sequences for the silicone. In these sequences, the silicone exhibits hypersignal while there is suppression of the water signal, the reverse being found in the sequences with silicone suppression. The acquisition should be performed in at least two orthogonal planes, in

order to allow a better distinction between complex radiating folds and early intracapsular ruptures. The administration of contrast is not necessary if the study is only directed to the evaluation of the integrity of the implants.²⁰

For the correct interpretation of the MRI study, knowledge of the usual aspect of implants and findings suggestive of complications is fundamental. To avoid potential diagnostic errors, the type of surgical approach and its date should be known for a better interpretation of early complications, as well as the existence or not of previous revision justifying the presence of free silicone related to previous ruptures (Figure 6).¹³

The evaluation implants by MR is contemplated in the last edition of BIRADS (5th edition), allowing a systematized evaluation and the standardization of the lexicon used.

The first step is to determine the type of implant (single vs double lumen) and its filling (silicone vs. saline). This latter distinction can be made in the T2-weighted sequences, since the saline implants exhibit hypersignal labeling, while the silicone imprints exhibit intermediate signal. In saline implants, a valve is usually present, usually located in subareolar topography. It is important not to confuse this

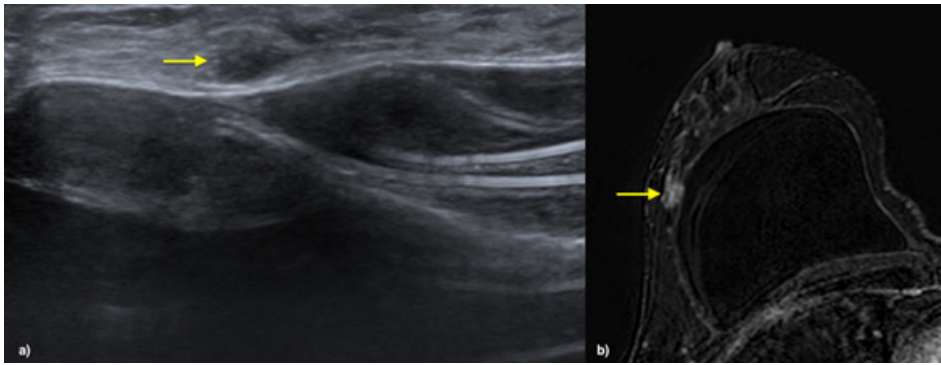


Figure 16 – The same ultrasound study (Figure 15) also revealed the presence of a nodule with indeterminate ultrasound characteristics, and a complementary MRI study was performed. In the study of MR (b), this presented enhancement after administration of contrast, and a directed biopsy was performed, which revealed that it was a foreign body granuloma.

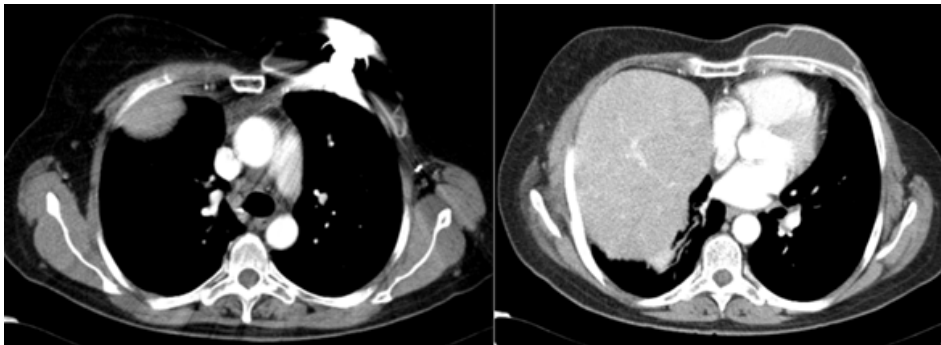


Figure 17 – A 47-year-old woman with a history of left mastectomy, showing an expander due to the presence of multiple artifacts at the level of her valve. The MRI study is contraindicated in some expanders, and its model should always be known before each exam. (<http://www.mrisafety.com/>).

valve with the markers used for its orientation during the surgical procedure (Figure 18).

Next, the location of the implants (retro-glandular vs. retro-pectoral) should be identified.

Its remaining evaluation includes its usual (oval) morphology, as an implant with rounded morphology may be a sign of capsular contracture.

The evaluation of its contours, defined externally by the fibrous capsule, is represented by a hyposignal-labeled line in T2. The presence of a small amount of peri-prosthetic fluid is common, especially in textured implants, resulting from the local inflammatory reaction to the foreign body.

The different types of irregularity of the contour must be distinguished from each other, since they may reflect quite different alterations. Radial folds are a rather frequent finding, translated by the presence of a perpendicular invagination of the inner membrane into the prosthesis, where the existence of an implant without at least one of these folds is very rare.



Figure 18 – MRI of 47-year-old woman submitted to left mastectomy in 2015, with symmetrization mammoplasty and placement of bilateral implants. In this sequence (T2) it is possible to verify the intermediate signal of the silicone of the implants and the presence of orientation markers (arrows).

These can be simple or complex, being one of the main pitfalls in the evaluation of eventual intracapsular rupture (Figure 19).

The undulations of the prosthetic contour are a finding with no pathological significance, resulting from the process of adapting a malleable implant to the surgical loca. Radial folds, as well as the presence of a small amount of peri-prosthetic fluid and contour curls constitute alterations in the normal spectrum of mammary implants (Figure 20).¹²

The presence of small protrusions or focal herniations are readily identified, and although do not diagnose rupture, they often occur simultaneously, as they translate areas of fragility of the fibrous capsule.

The presence of intracapsular rupture is more easily detected through this imaging method. The most reliable finding of this complication is the “linguini signal”, described by Gorczyca et al., in 1992, with a sensitivity and specificity of 96% and 77% respectively.²¹ This signal consists of a late stage of this type of rupture, associated with partial or total collapse of the elastomer shell. Imaging is represented by the presence of multiple hypointense curvilinear lines within the hyperintense contents of the implant (Figures 12 and 21).

As described in the ultrasound, there are two findings that represent earlier stages of this type of rupture, namely the locking signal (earlier) in which there is no associated collapse and the signal of the subcapsular (intermediate) line in which there may be minimal collapse of the casing. The “lock” results from the extravasation of silicone by the apex of a radiating fold, causing expansion of this region, which will later migrate into the intracapsular space, with a detachment of the inner membrane and forming the subcapsular line. The presence of some T2-weighted hypersignal foci inside the implant (“salad oil sign”) were also described as a sign suggesting this type of complication, although not a reliable signal as it may result from the injection of corticosteroids or antiseptics during the surgical procedure (Figure 22).

Extracapsular rupture may be associated with any degree of collapse, and the search for free silicone in topography

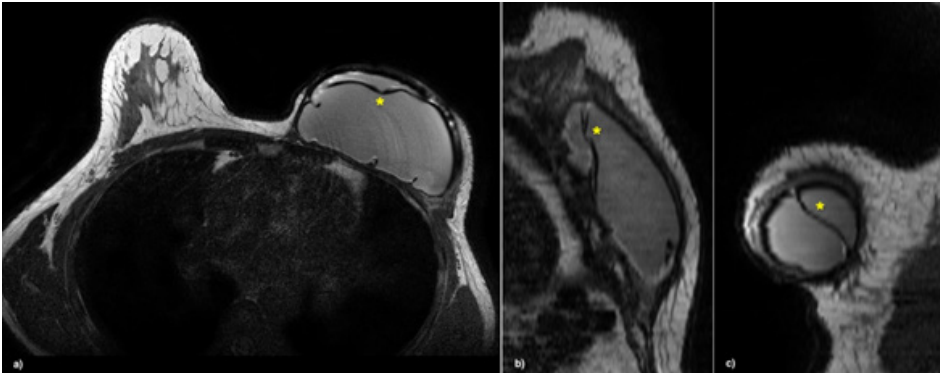


Figure 19 – A 38-year-old female with left mastectomy and reconstruction with a *Latissimus dorsi* flap and implants. The evaluation of the MRI study demonstrated the presence of what appeared to be the “sign of the subcapsular line” suggesting intracapsular rupture in the axial plane (a). Its evaluation in the other orthogonal planes (coronal (b) and sagittal (c)) revealed a pitfall, conditioned by the presence of a complex radiating fold.

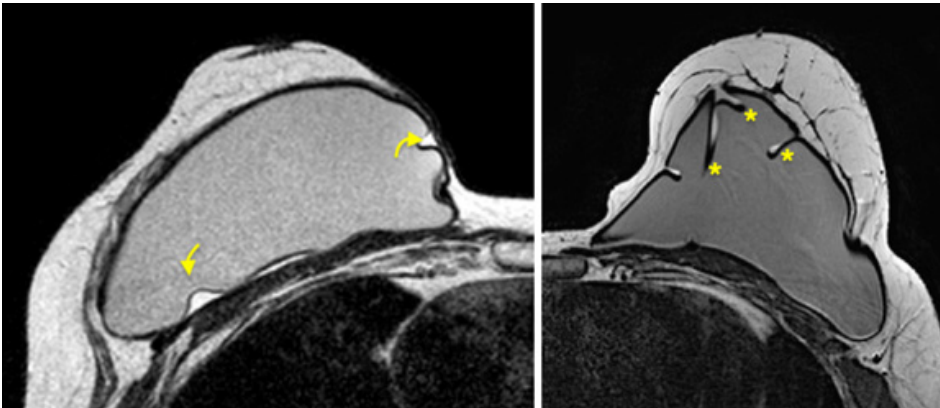


Figure 20 – Changes in the normal spectrum of mammary implants in the MRI study (T2-weighted sequence), namely the presence of undulations and minimal amount of periosthetic fluid (arrows) and the presence of radiating folds (asterisks).

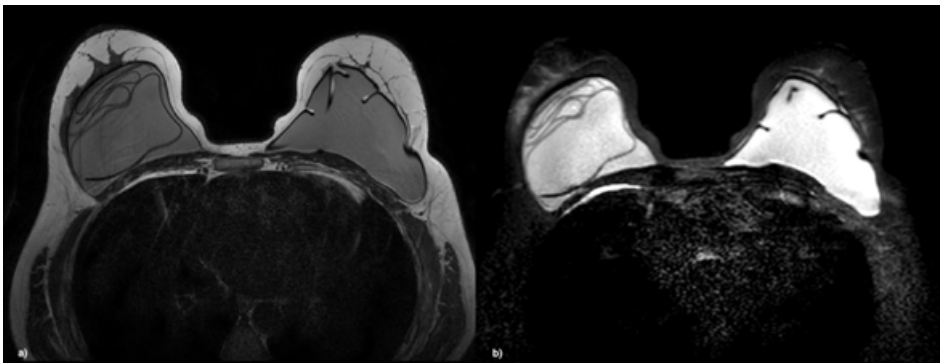


Figure 21 – MRI study revealing the presence of the “Linguini signal”, diagnosis of intracapsular rupture, evidenced in T2 (a) and selective sequence for silicone (b).

outside the fibrous capsule should be performed whenever an intracapsular rupture is detected. In MRI this is translated by the presence of foci with a signal identical to that of silicon in the sequences dedicated to its evaluation. When confluent, they may form silicone granulomas, which, like malignant lesions, may undergo growth and display enhancement after administration of contrast, being practically indistinguishable only by imaging techniques. In these cases, a directed ultrasound study should be performed in an attempt to identify the snowstorm signal, and in the absence of this, a biopsy should be performed. As with other imaging techniques, lymph nodes may also exhibit the same signal as silicone, but are not pathognomonic of extracapsular rupture (Figure 23).¹⁴

4.2.3 Adenomegalies in the internal mammary chain

The presence of adenomegaly in the internal mammary chain is a diagnostic challenge, especially in women with prostheses after oncoplastic surgery. These may result from a non-specific inflammatory process, translate the presence of extracapsular rupture or a lymph node metastasis. A study (Sutton et al) demonstrated the presence of adenomegaly in



Figure 22 – An 85-year-old woman with bilateral mastectomy due to bilateral carcinoma, with reconstruction with implants 20 years ago. The MRI study revealed the presence of bilaterally intracapsular rupture signs, to the right, translated by the presence of the “subcapsular line” sign (arrow) and left by the “linguini” and “salad oil” signs (arrow and asterisk respectively).

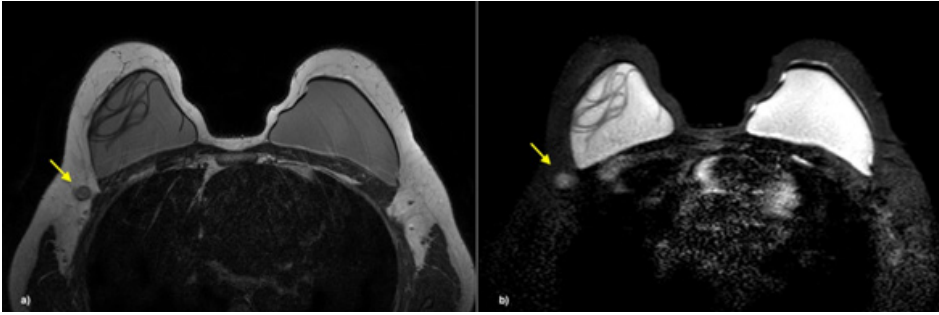


Figure 23 – MRI study of a woman with right intracapsular rupture (“linguini” sign), identifying axillary adenomegaly (arrow) in T2-weighted sequence (a), which shows hypersignal in the selective sequence of silicone (b) indicating coexistence of extracapsular rupture.

this lymphatic chain in up to one third of women with silicone implants. Of the 207 women with adenomegalies studied, only one of them revealed metastatic adenopathy. Thus, this finding should be classified as BIRADS 3, guaranteeing a short-term evaluation for 2 years, in detriment of the biopsy (Figure 24).^{6,22}

4.2.4 Large cell anaplastic lymphoma

It is a rare late complication, recently described in the literature, with an incidence between 1: 500 and 1: 3,000,000 in women with implants.²³ Its diagnosis is made, on average, 10 years after surgery. The systemic symptoms are rare and the clinic is non-specific. This entity usually translates into a peri-prosthetic effusion and / or mass, which can progressively progress to soft tissue injury. Ultrasound and MRI are the most sensitive imaging methods for its detection. Thus, the presence of “new” peri-prosthetic effusion after the postoperative period (1 year) should alert the radiologist to its diagnosis, suggesting the aspiration of the effusion and its laboratory analysis with flow cytometry.⁶

4.2.5 Complications related to reconstruction

Several types of complications may arise after oncoplastic surgery, their incidence being related to the timing of

chemotherapy and / or radiotherapy, as well as to the type of reconstruction. The early complications are similar to those observed after augmentation mammoplasty, adding cutaneous necrosis and dehiscence of the suture.²⁴ A complication frequently related to TRAM is fat necrosis, occurring in 15% of cases. Other complications include total or partial necrosis of the flap (0.3% and 2-6%) and abdominal wall herniation (Figure 25 and 26).⁶

4.2.6 Relapse / Surveillance

Breast implants are not related to the increased risk of breast cancer, however they may reduce the acuity of the usual screening methods.¹² Specific mammographic incidents such as Eklund, with removal of the prosthesis and isolated compression of the mammary parenchyma, aim to increase sensitivity in the detection of lesions in women with implants. MRI is, however, the best technique for its detection, explaining the fact that the MRI protocol in women with implants includes in most cases the dynamic post-contrast study, in order to increase its detection rate (Figures 27 and 28).

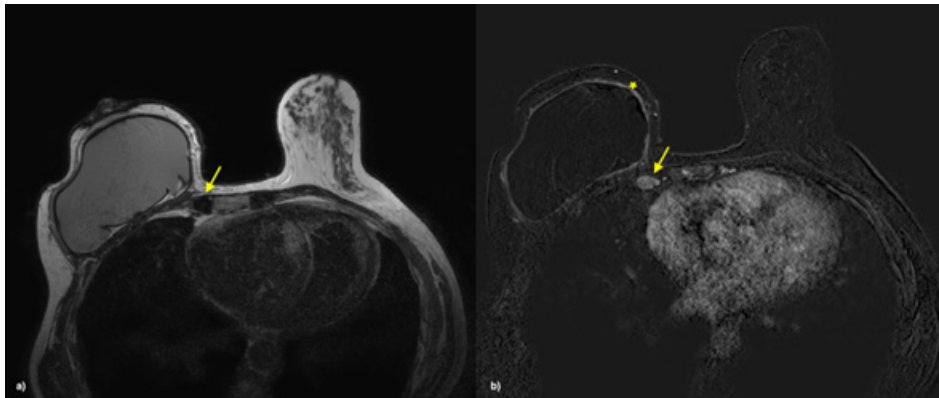


Figure 24 – A 46-year-old woman with a history of right-sided breast cancer with reconstruction with breast implant. The MRI study revealed the presence of adenomegaly in the right internal mammary chain (arrow), evidenced in the weighted sequence in T2 (a) and T1 after contrast (b). Concomitantly, there is an early and diffuse enhancement of the fibrous capsule (asterisk), in a probable relation with the underlying inflammatory process, thus explaining the reactive nature of the identified adenomegaly.

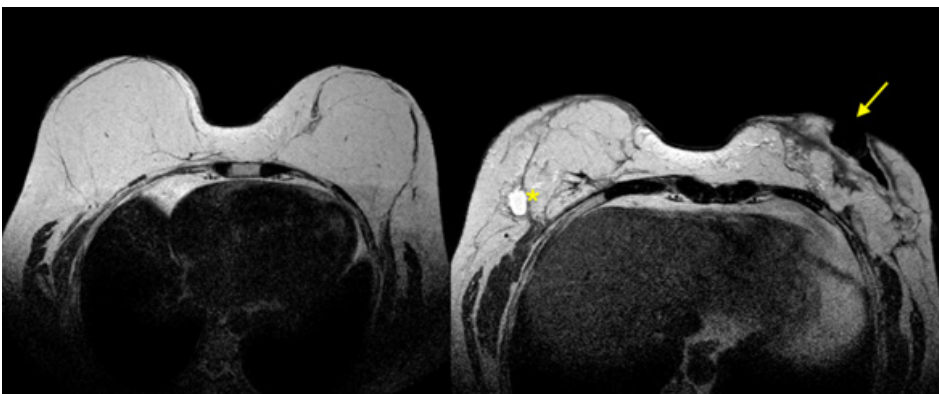


Figure 25 – A 55-year-old woman with a history of oncoplastic surgery with rectus abdominis muscle flap (TRAM). The MRI study revealed the presence of a small seroma on the right (asterisk) and the presence of cutaneous necrosis and dehiscence of the left suture (arrow).

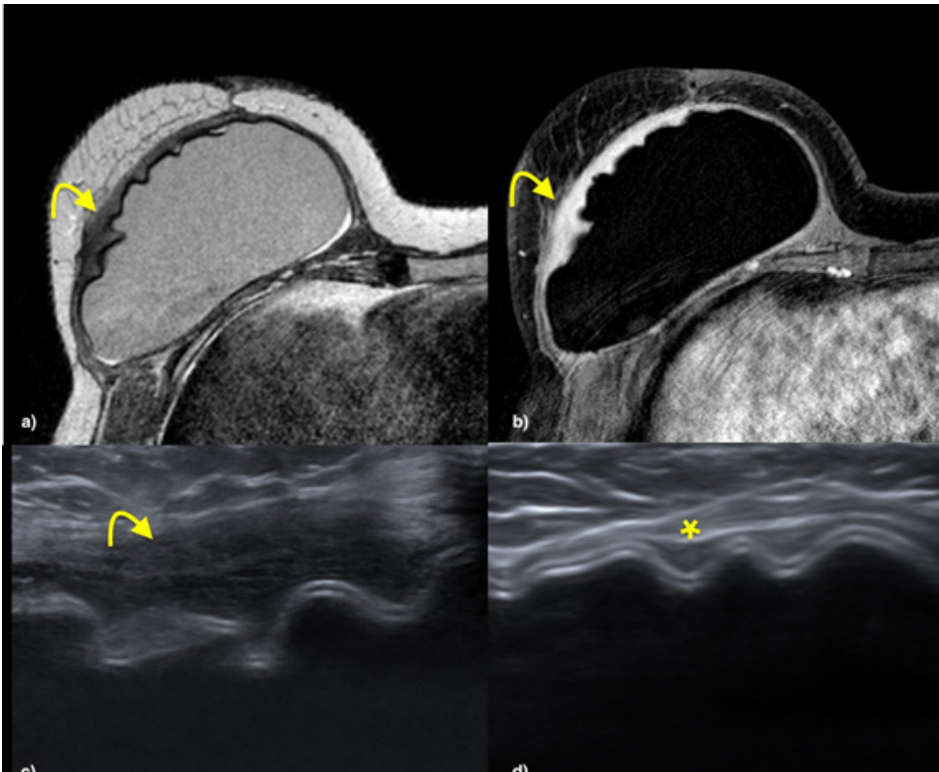


Figure 26 – A 42-year-old female with a history of breast carcinoma on the right with breast reconstruction with a *Latissimus dorsi* flap and implant. The MRI study demonstrated marked thickening of the muscle flap (a), with diffuse enhancement after administration of contrast (b) in relation to the local inflammatory process. The complementary ultrasound study (c) was concurrent, and a favorable clinical course was observed after antibiotic therapy and anti-inflammatory therapy, with disappearance of these findings in the ultrasound of control at 3 months (d).

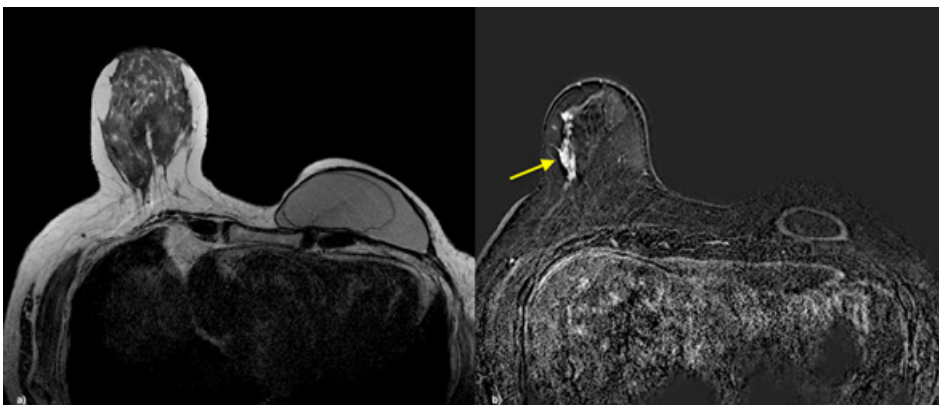


Figure 27 – A 71-year-old woman with a history of breast carcinoma on the left with breast reconstruction with a *Latissimus dorsi* flap and implant. In the MRI study, the presence of intracapsular rupture on the right was verified, identifying the signal of the subcapsular line (a). In the dynamic study after administration of contrast (b), a non-mass enhancement of the linear type was identified in the inferolateral quadrant of the contralateral breast, which after biopsy revealed a ductal carcinoma in situ.

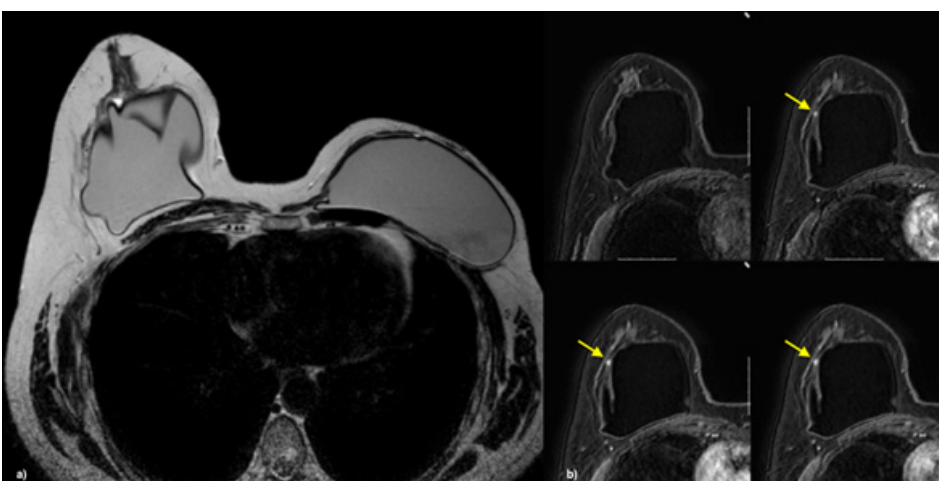


Figure 28 – Follow-up MRI in a left mastectomized woman with reconstruction with bilateral implants. Assessment of T2-weighted sequences (a) revealed no signs of prosthesis-related complications. However, in the post-contrast (b) dynamic study, the presence of a focus of enhancement was verified, which after a second-look ultrasound study and echoguided micro biopsy, an invasive ductal carcinoma was found.

5. Conclusion

With the exponential increase of women with breast implants in the last decades, it is therefore imperative to know the main associated complications and the advantages and limitations of each of the imaging techniques. Mammography has a limited role in its evaluation, and only the diagnosis of extracapsular rupture is possible. Ultrasonography allows a more detailed evaluation of the implant, namely regarding the possibility of intracapsular rupture. However, MRI is the technique with greater sensitivity and specificity in the detection and characterization of these complications. Thus,

given its nonspecific clinic, Radiology plays a central role in its monitoring, allowing early detection and better diagnostic and therapeutic guidance.

Abbreviations:

USA – United States of America
TRAM - Transverse rectus abdominis myocutaneous
DIEP - Deep inferior epigastric perforators
MR – Magnetic resonance
FDA - Food and Drug Administration
DGH – Directorate General for Health
CT - Computerized Tomography

Received /Recebido 13/12/2017

Acceptance / Aceite 02/02/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Supporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Czerny V. Plastic replacement of the breast with a lipoma. *Chir Kong Verhandel.* 1895;2:216.
2. Cronin TD, Brauer RO. Augmentation mammoplasty. *Surg Clin North Am.* 1971;51:441-52.
3. International Society of Plastic Aesthetic Surgery. (2017) International Study On Aesthetic/Cosmetic Surgery Procedures Performed In 2016. Available at: <http://www.isaps.org/Media/Default/Current%20News/GlobalStatistics2016.pdf>. Accessed November 04, 2017.
4. Albornoz CR, Bach PB, Mehrara BJ, Disa JJ, Pusic AL, McCarthy CM, et al. A paradigm shift in U.S. breast reconstruction: increasing implant rates. *Plast Reconstr Surg.* 2013;131:15-23.
5. Pelosi, Marco A. III, Pelosi, Marco A. II. Breast augmentation. *Obstet Gynecol Clin North Am.* 2010;37:533-46.
6. Green L.A, Karow JA, Toman JE, Lostumbo A, Xie K. Review of breast augmentation and reconstruction for the radiologist with emphasis on MRI. *Clinical Imaging.* 2018;47:101-17.
7. Maxwell GP, Gabriel A. Breast implant design. *Gland Surg.* 2017;6:148-53.
8. Yang N, Muradali D. The augmented breast: a pictorial review of the abnormal and unusual. *AJR Am J Roentgenol.* 2011;196:451-60
9. Pittet B, Montandon D, Pittet D. Infection in breast implants. *Lancet Infect Dis.* 2005;5:94-106.

10. Namnoum J, Largent J, Kaplan H, et al. Primary breast augmentation clinical trial outcomes stratified by surgical incision, anatomical placement and implant device type. *J Plast Reconstr Aesthet Surg.* 2013;66:1165-72.
11. Siggelkow W, Faridi A, Spiritus K, Klinge U, Rath W, Klosterhalfen B. Histological analysis of silicone breast implant capsules and correlation with capsular contracture. *Biomaterials.* 2003;24:1101-09.
12. Juanpere S, Perez E, Huc O, Motos N, Pont J, Pedraza S. Imaging of breast implants - a pictorial review. *Insights into Imaging.* 2011;2:653-70.
13. Gorczyca David P, Gorczyca Stephanie M, Gorczyca Kathryn. The diagnosis of silicone breast implant rupture. *Plast Reconstr Surg.* 2007;120:49-61.
14. Seiler SJ, Sharma PB, Hayes JC, Ganti R, Mootz AR, Eads ED, Teotia SS, Evans WP. Multimodality imaging-based evaluation of single-lumen silicone breast implants for rupture. *Radiographics.* 2017;37:366-82.
15. Brenner RJ. Evaluation of breast silicone implants. *Magn Reson Imaging Clin N Am.* 2013;21:547-60.
16. Norma da DGS n° 051/2011, “Abordagem Imagiológica da Mama Feminina”.
17. Di Benedetto G, Cecchini S, Grasseti L, Baldassarre S, Valeri G, Leva L, et al. Comparative study of breast implant rupture using mammography, sonography, and magnetic resonance imaging: correlation with surgical findings. *Breast J.* 2008;14:532-7
18. Everson LI, Parantainen H, Detlie T, Stillman AE, Olson PN, Landis G, et al. Diagnosis of breast implant rupture: imaging findings and relative efficacies of imaging techniques. *AJR Am J Roentgenol.* 1994;163:57-60.
19. Berg WA, Caskey CI, Hamper UM, Anderson ND, Chang BW, Sheth S, et al. Diagnosing breast implant rupture with MR imaging, US, and mammography. *RadioGraphics* 1993;13:1323-36
20. JF Wiedenhoefer, H Shahid, C Dornbluth, P Otto, K Kist. MR imaging of breast implants: useful information for the interpreting radiologist. *Appl Radiol.* 2015;44:18-24.
21. Gorczyca DP, Sinha S, Ahn CY, et al. Silicone breast implants in vivo: MR imaging. *Radiology.* 1992;185:407-10.
22. Sutton EJ, Watson EJ, Gibbons G, Goldman DA, Moskowitz CS, Jochelson MS, et al. Incidence of internal mammary lymph nodes with silicone breast implants at MR imaging after oncologic surgery. *Radiology.* 2015;277:381-87.
23. Ye X, Shokrollahi K, Rozen WM, Conyers R, Wright P, Kenner L, et al. Anaplastic large cell lymphoma (ALCL) and breast implants: breaking down the evidence. *Mutat. Res./Rev. Mutat. Res.* 2014;762:123-32.
24. Ilonzo N, Tsang A, Tsantes S, Estabrook A, Ma AMT. Breast reconstruction after mastectomy: a ten-year analysis of trends and immediate postoperative outcomes. *Breast.* 2016;32:7-12.

Computed Tomography in the Evaluation of Lung Transplant Chronic Rejection

Tomografia Computorizada na Avaliação da Rejeição Crónica nos Transplantes Pulmonares

João Lopes, Marta Simões

Serviço de Radiologia, Hospital de Santa Marta,
Centro Hospitalar Lisboa Central, Lisboa,
Portugal

Address

João Lopes
Serviço de Radiologia
Hospital de Santa Marta, Centro Hospitalar
Lisboa Central
Rua de Santa Marta 50
1169-024, Lisboa, Portugal
e-mail: jpa.lopes@gmail.com

Abstract

Lung transplantation is an increasingly common therapeutic option in end-stage pulmonary diseases. One of the main causes of medium and long-term graft failure is chronic rejection, clinically represented by bronchiolitis obliterans syndrome. The early diagnosis of chronic rejection allows optimization of immunosuppressive therapy in order to delay its progression.

In this paper, we review and illustrate the characteristics of chronic lung rejection in high-resolution computed tomography to promote its early diagnosis in follow-up examinations.

At an early stage, during the first year after transplantation, subtle features such as reduction of peripheral bronchovascular markers, thickening of the septal lines, and decreased lung volumes may suggest the diagnosis even before clinical changes appear. Mid-term features are represented by bronchiectasis and bronchial wall thickening, and present low sensitivity, but high specificity in the diagnosis of chronic rejection. Its appearance occurs simultaneously with the clinical diagnosis of bronchiolitis obliterans syndrome. Lung attenuation abnormalities appear in late stages of the disease. Air trapping is related with small airway obstruction and mosaic attenuation pattern with ventilation-perfusion mismatch. Fibrotic changes of the lung parenchyma characterize advanced stages of chronic graft rejection, leading to important functional repercussion.

High-resolution computed tomography has helped to overcome the limitations of clinical criteria in the diagnosis of obliterans bronchiolitis syndrome and promoted an earlier diagnosis of chronic rejection after lung transplantation.

Keywords

CT; Lung transplantation; Transplant rejection; Bronchiolitis obliterans.

Resumo

O transplante pulmonar é uma opção terapêutica cada vez mais frequente em doenças pulmonares em estágio terminal. Uma das principais causas de falência do enxerto, a médio e longo prazo, é a rejeição crónica, traduzindo-se sob a forma de síndrome de bronquiolite obliterante. O diagnóstico precoce desta entidade permite otimizar a terapêutica imunossupressora, limitando a sua progressão.

Este trabalho reúne, discute e ilustra as características da rejeição crónica dos enxertos pulmonares na tomografia computadorizada de alta resolução, com o objetivo de facilitar o seu reconhecimento em exames de seguimento e, promover o diagnóstico precoce desta entidade.

Numa fase precoce, durante o primeiro ano após o transplante, alterações subtis como a redução das marcas broncovasculares periféricas, o espessamento das linhas septais e a diminuição do volume pulmonar podem indiciar o seu diagnóstico, mesmo antes do aparecimento de alterações clínicas. A médio prazo, aquando do diagnóstico clínico, podem observar-se bronquiectasias e espessamento brônquico, características com baixa sensibilidade, mas de alta especificidade no diagnóstico da rejeição crónica. As alterações da atenuação pulmonar surgem em fases mais avançadas desta síndrome, evidenciando padrão de retenção aérea por obstrução das pequenas vias respiratórias, associado a padrão de atenuação em mosaico, condicionado por alterações da ventilação-perfusão. Tardamente, a rejeição crónica do enxerto caracteriza-se por alterações fibróticas do parênquima pulmonar com importante repercussão funcional.

A tomografia computadorizada de alta resolução tem ajudado a ultrapassar as limitações dos critérios clínicos no diagnóstico da síndrome de bronquiolite obliterante e promovido o diagnóstico mais precoce da rejeição crónica dos enxertos pulmonares.

Palavras-chave

TC; Transplante pulmonar; Rejeição de transplante; Bronquiolite obliterante.

Introduction

Pulmonary transplantation is currently a therapeutic option that promotes increased survival and quality of life in patients

with end-stage pulmonary diseases who meet criteria for indication of this intervention.¹

Complications vary according to temporal progression, the infectious interurrences being the main cause of mortality

in the first six months after transplantation and chronic graft rejection the main cause of mortality after that period.² From the histopathological point of view, chronic rejection is characterized by proliferative submucosal fibrotic changes that condition obstruction of the small airways, a process known as obliterating bronchiolitis, which can reach up to 50% of transplanted patients.³

Chronic rejection has strong associations established with previous episodes of acute rejection and cytomegalovirus infection. Clinically, it may be manifested by progressive worsening cough and dyspnea, associated with decreased FEV1-dependent on the pathology of small airways.⁴

The concept of bronchiolitis obliterans syndrome, defined as the $\geq 20\%$ reduction in FEV1 in relation to the baseline value after transplantation, was created by the International Society for Heart and Lung Transplantation due to the low sensitivity of lung biopsies in the diagnostic confirmation of chronic rejection of the pulmonary graft.⁵ In the differential diagnosis, other causes justifying FEV1 reduction, such as infectious interurrences or episodes of acute rejection of the graft, should be excluded. This functional evaluation, however, presents some limitations, especially in patients with uni-pulmonary transplants, in which the lung function of the native lung can influence the results obtained.³

Early diagnosis of chronic rejection of lung transplantation is essential, since adjustment of immunosuppressive therapy may delay its progression and significantly improve prognosis. Given the limitations associated with clinical and histopathological evaluation, imaging methods, namely computerized tomography, have gained prominence in the search for early signs of chronic rejection.^{6,7}

This review paper gathers, discusses and exemplifies the tomographic semiology known to date on chronic rejection in lung transplants, seeking to simplify the approach of the radiologist when confronted with this situation in his clinical practice.

Imaging findings

Imaging of pulmonary grafts is performed using X-ray and computed tomography scans of serial thorax, and radiographic findings of bronchiolitis obliterans, which usually only appear six months after transplantation, are non-specific. They usually consist of segmental atelectasis foci, decrease in peripheral vascular markers and pulmonary volume.⁴ However, lung parenchyma and bronchial structures ability to be appreciated is much higher in computed tomography, allowing greater sensitivity in the detection and characterization of chronic alterations.^{3,8}

The onset of bronchiolitis obliterans follows a chronological sequence of changes in computed tomography, which can be organized in early, mid-term and late alterations and are characterized hereafter.³

Early changes

It is possible to recognize imaging changes suggesting chronic rejection even before the clinical diagnosis of bronchiolitis obliterans syndrome. Among these aspects are the reduction of the peripheral bronchovascular markers (Fig. 1), the thickening of the septal lines (Fig. 2) and the reduction of the volume of the transplanted lung (Fig. 3). These findings usually occur during the first year after transplantation, however, they have limited diagnostic utility

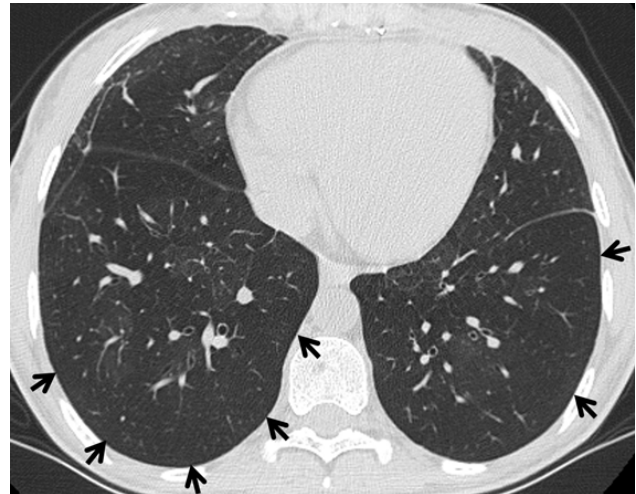


Figure 1 - A 27-year-old woman with bronchiolitis obliterans syndrome after bi-pulmonary transplantation. In this high-resolution axial image obtained one year after transplantation, the bronchovascular markers are reduced to the periphery, affecting mainly the posterior slope of the lower lobes. Additionally there is discrete pattern of mosaic attenuation.

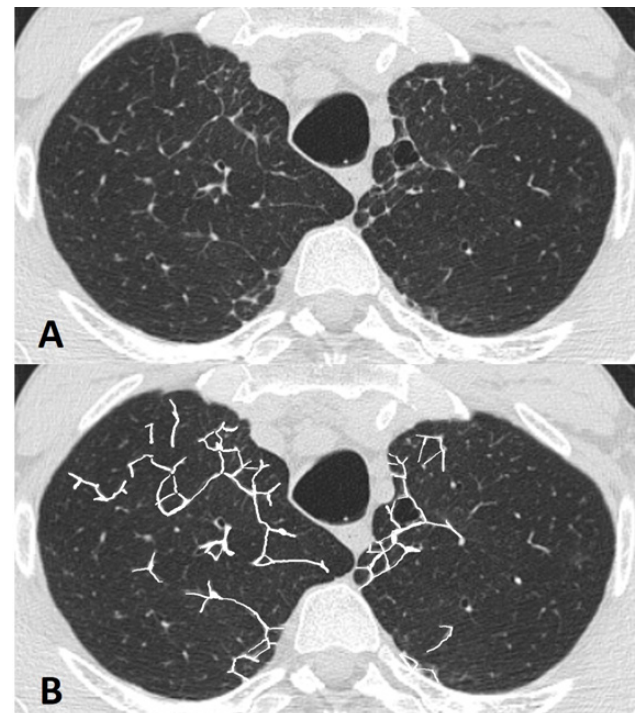


Figure 2 - Thickening of septal lines in a 37-year-old man with bronchiolitis obliterans syndrome after bi-pulmonary transplantation. High resolution axial image (A) obtained 5 and a half years after transplantation, where the acinar anatomy is evidenced due to the thickening of the intralobular and interlobular septa. These changes are illustrated in B.

in the prediction of chronic rejection due to their subtle and inconstant nature.³

Medium-term changes

Bronchiectasis (Fig. 4) accompanies the clinical diagnosis of the bronchiolitis obliterans syndrome and is defined as the presence of bronchial dilations with a bronchial-artery ratio greater than 1 or the visualization of the bronchial lumen within 1 cm of the costal pleura.⁹

Increased resistance in the small airways caused by obliteration of small bronchial structures is one of the accepted etiologic mechanisms in the formation of bronchiectasis in these

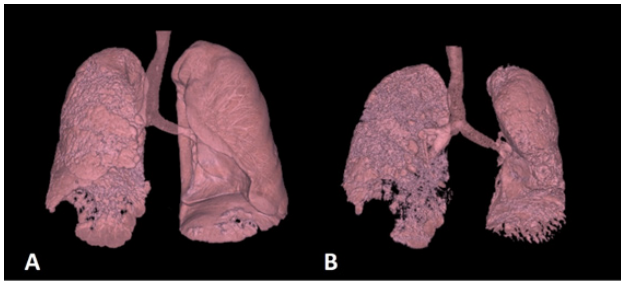


Figure 3 - Three-dimensional reconstructions (volume rendering technique) in men submitted to left uni-pulmonary transplantation, 4 months (A) and 4 years (B) after transplantation. Between the two evaluations there is marked volumetric reduction of the graft resulting from chronic rejection. There is also a volumetric reduction of the native right lung, although to a lesser degree, probably due to the progression of the base scaly interstitial pneumonia.

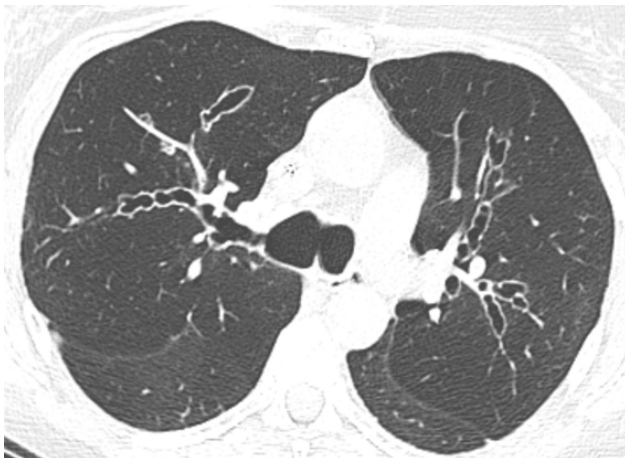


Figure 4 - Bilateral variceal bronchiectasis in a woman with bronchiolitis obliterans syndrome, after bilateral lung transplantation, one and a half years before the examination. Note the associated bronchial thickening.

patients. However, frequent acute infectious complications in lung transplant patients are also a known cause of bronchiectasis, which is not related to chronic graft rejection.³ Thickening of the bronchial wall (Fig. 5) is another aspect frequently observed and often associated with bronchiectasis, and its evaluation is performed subjectively by the radiologist.⁹ Both bronchiectasis and thickening of the bronchial wall have low sensitivity but high specificity in the diagnosis of chronic rejection of the pulmonary graft.^{7,9}

Late changes

Changes in normal pulmonary attenuation, such as the presence of air trapping (Fig. 6) or mosaic attenuation pattern (Fig. 7) which typically appear in late phases of chronic rejection, are often associated with advanced stages of the disease and caused by bronchial obliteration and small airway obstruction.³

Air trapping

The attenuation of the lung parenchyma depends on the amount of intrapulmonary air, decreasing in inspiration and increasing in the exhalation.

The presence of air trapping consists in the existence of areas of pulmonary parenchyma in which the increase of attenuation with expiration is smaller than it would be expected. In affected areas, the expected volume reduction between inspiratory and expiratory acquisition is also lower than normal.^{10,11}



Figure 5 - Bronchial thickening (arrows) in a man with bronchiolitis obliterans syndrome, submitted six years before to bilateral lung transplantation.

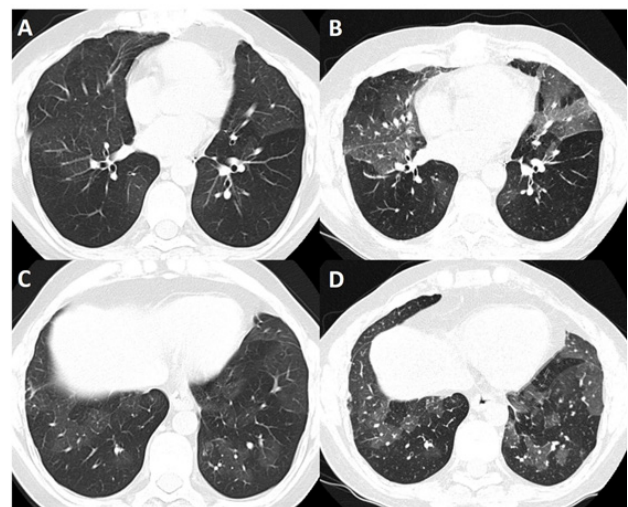


Figure 6 - Examples of air trapping in two different segments, in a man with bronchiolitis obliterans syndrome three years after bi-pulmonary transplantation. Changes in pulmonary parenchymal attenuation are tenuous on inspiration acquisition (A, C). At the end of the expiration air trapping becomes evidently marked (B, D). The areas with the highest density correspond to areas of normal attenuation.

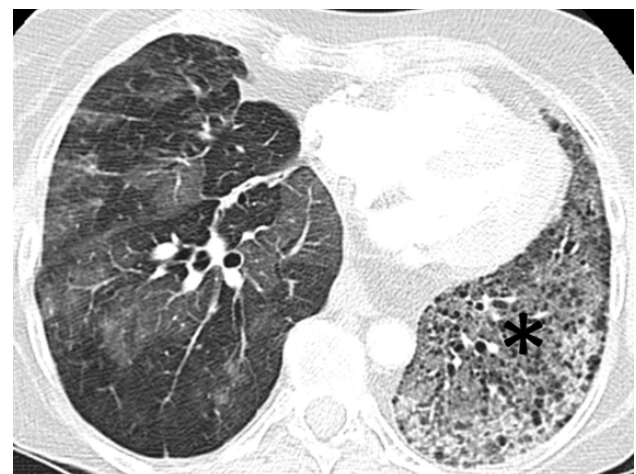


Figure 7 - Acquisition in inspiration showing mosaic attenuation pattern in a woman with bronchiolitis obliterans syndrome four years after right unilateral lung transplantation. Native left lung with advanced stage changes due to extrinsic allergic alveolitis (*).

In clinical practice, the air trapping evaluation is usually qualitative. Several studies have been developed to eliminate the subjectivity of this type of evaluation and to find useful semiquantitative and quantitative measures. Among these studies, Bankier et al determined the 33% parenchyma limit affected by air trapping as the value from which computed tomography allows the diagnosis of bronchiolitis obliterans syndrome to be predicted in a sensitive, specific and precise manner, even before clinical appearance. Knollmann et al showed that it is possible to use quantitative measures to predict the diagnosis of bronchiolitis obliterans syndrome using a spirometric gating method, but the complexity of this technique has limited its application to current clinical practice.

Notes and technical aspects useful in evaluating air trapping:

- 1) The attenuation of the pulmonary parenchyma decreases from the bases to the apex of the lungs and is more homogeneous in inspiration.¹⁰
- 2) During exhalation, it is possible to observe some areas of air trapping in healthy individuals, especially in locations adjacent to the small fissure or in the apical segments of the lower lobes.^{9,11}
- 3) In an individual assessment over time, air trapping is an inconstant finding in a significant number of patients.⁹
- 4) For proper detection of air trapping it is essential to compare acquisitions in inspiration and expiration.⁶
- 5) It is not recommended to limit expiratory acquisition to some images in predefined plans as it is usual in the study of other pulmonary pathologies. The heterogeneous distribution of the changes related to the bronchiolitis obliterans syndrome necessitates the study of the full extent of the pulmonary parenchyma, even at expiration.
- 6) Prior patient training of breath-hold at maximal inspiratory and expiratory positions contributes to acquisition at appropriate ventilatory times and reduces movement artifacts.

Mosaic Attenuation Pattern

The mosaic attenuation is defined by the presence of areas of low attenuation of the pulmonary parenchyma intercalated by areas of normal attenuation, in the acquisition in inspiration. The areas of lower attenuation result from decreased vascularization due to ventilation-perfusion changes.¹⁰

The mosaic attenuation pattern has low sensitivity but high specificity in the diagnosis of bronchiolitis obliterans syndrome, similar to that observed in the presence of bronchiectasis or thickening of the bronchial wall.⁹

Graft fibrosis

With the progression of the bronchiolitis obliterans syndrome to the terminal stages of the disease, aspects

such as thickening of the septal lines, loss of lung volume or bronchiectasis culminate in irreversible fibrotic changes (Fig. 8).

When the fibrotic alterations are focused on the upper lobes, they constitute a distinct rare entity associated with chronic pulmonary rejection, which arises 1 to 4 years after transplantation and is characterized by thickening of the interlobular septa, reticular and depolarized glass opacities, traction bronchiectasis, honeycomb alterations, architectural distortion and loss of lung volume.^{1,2}

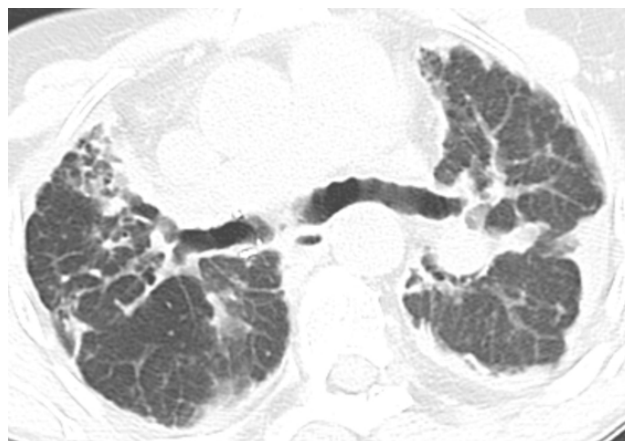


Figure 8 - End-stage chronic lung rejection in a 47-year-old woman with bronchiolitis obliterans syndrome, two years after bilateral lung transplantation. In the image, coarse crosslinking is evident, associated with architectural distortion, traction bronchiectasis and significant loss of lung volume

Conclusion

The radiologist plays a leading role in the evaluation of patients undergoing lung transplantation and should be familiar with the temporal progression of the main imaging findings related to chronic rejection.

In the evaluation of the transplanted pulmonary patient, parenchymal alterations related to the reduction of the peripheral bronchovascular markers, thickening of the septal lines, loss of volume, bronchiectasis, bronchial wall thickening, air trapping or pattern of mosaic attenuation should make the hypothesis of bronchiolitis obliterans syndrome be considered. Fibrotic changes of the pulmonary parenchyma are of relevance in the advanced stages of chronic rejection.

Received / Recebido 02/11/2017

Acceptance / Aceite 11/01/2018

Divulgações Éticas / Ethical disclosures

Conflicts of interest: The authors have no conflicts of interest to declare.
Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Ng YL, Paul N, Patsios D, Walsham A, Chung T-B, Keshavjee S, et al. Imaging of lung transplantation: review. *AJR Am J Roentgenol.* 2009;192: S1-13.
2. Krishnam MS, Suh RD, Tomasian A, Goldin JG, Lai C, Brown K, et al. Postoperative complications of lung transplantation: radiologic findings along a time continuum. *Radiographics.* 2007;27:957-74.
3. Ikonen T, Kivisaari L, Taskinen E, Piilonen A, Harjula AL. High-resolution CT in long-term follow-up after lung transplantation. *Chest.* 1997;111:370-6.
4. Murray JG, McAdams HP, Erasmus JJ, Patz EF, Tapson V. Complications of lung transplantation: Radiologic findings. *Am J Roentgenol.* 1996;166:1405-11.
5. Cooper JD, Billingham M, Egan T, Hertz MI, Higenbottam T, Lynch J, et al. A working formulation for the standardization of nomenclature and for clinical staging of chronic dysfunction in lung allografts. International Society for Heart and Lung Transplantation. *J Hear Lung Transpl.* 1993;12:713-6.
6. Siegel MJ, Bhalla S, Gutierrez FR, Hildebolt C, Sweet S. Post-lung transplantation bronchiolitis obliterans syndrome: usefulness of expiratory thin-section CT for diagnosis. *Radiology.* 2001;220:455-62.
7. Leung AN, Fisher K, Valentine V, Girgis RE, Berry GJ, Robbins RC, et al. Bronchiolitis obliterans after lung transplantation: Detection using expiratory HRCT. *Chest.* 1998;113:365-670.
8. Worthy SA, Park CS, Kim JS, Muller NL. Bronchiolitis obliterans after lung transplantation: high-resolution CT findings in 15 patients. *AJR Am J Roentgenol.* 1997;169:673-7.
9. Konec E, Gutierrez C, Chaparro C, Murray CP, Chung T, Crossin J, et al. Bronchiolitis obliterans syndrome in lung transplant recipients: can thin-section CT findings predict disease before its clinical appearance? *Radiology.* 2004;231:467-73.
10. Knollmann FD, Ewert R, Wünderlich T, Hetzer R, Felix R. Bronchiolitis obliterans syndrome in lung transplant recipients: use of spirometrically gated CT. *Radiology.* 2002;225:655-62.
11. Bankier AA, Van Muylem A, Knoop C, Estenne M, Gevenois PA. Bronchiolitis obliterans syndrome in heart-lung transplant recipients: diagnosis with expiratory CT. *Radiology.* 2001;218:533-9.

Images of Interest / Imagens de Interesse

Infarction of Torsed Lipomatous Appendage of the Falciform Ligament

Enfarte de apêndice lipomatoso torcido do ligamento falciforme

Carlos Francisco Silva, Vanessa Praxedes, Ana André

Centro Hospitalar de Setúbal, Serviço de
Imagiologia, Setúbal, Portugal

Address

Carlos Francisco Silva
Centro Hospitalar de Setúbal
Serviço de Radiologia
Rua Camilo Castelo Braco
2910-446 Setúbal, Portugal
email: carlos.f.silva@chs.min-saude.pt

Abstract

A 46-year-old man presented sudden onset of severe pain in the right upper quadrant. On abdominal examination, a well-defined pain and a subtle lump were obvious. Abdominal x-ray, ultrasound and laboratory tests were unremarkable. An abdominal CT scan was then requested, showing fat stranding and a torsed appearance of a lipomatous appendage of the falciform ligament, compatible with infarction. The pathophysiology of this entity is similar to the more commonly seen omental infarction and epiploic appendagitis, covering the spectrum of intra-abdominal focal fat infarction, recently described in literature.

Keywords

Infarction; Lipomatous appendage; Falciform ligament.

Resumo

Homem de 46 anos, apresentando forte dor súbita no hipocôndrio direito. Na avaliação abdominal evidenciava-se um ponto algíco e pequena tumefação bem definidos. Radiografia abdominal, ecografia e exames laboratoriais sem alterações dignas de registo. Foi então solicitada uma tomografia computadorizada abdominal, demonstrando sinais inflamatórios e aspetos de torção num apêndice lipomatoso do ligamento falciforme compatível com enfarte. A fisiopatologia desta entidade é semelhante ao enfarte omental mais comumente observado e à apendicite epiploica, abrangendo o espectro do enfarte focal de gordura intra-abdominal, recentemente descrito na literatura.

Palavras-chave

Enfarte; Apêndice lipomatoso; Ligamento falciforme.

A 46-year-old male presented sudden onset of severe pain in the right upper quadrant. On abdominal examination, a well-defined point of focal tenderness and a subtle lump were evident. Abdominal x-ray, ultrasound and laboratory tests were unremarkable. An abdominal CT scan was then requested, however, the report from an outsourced company suggested normal findings. After revision, the following morning, by a local attending radiologist and multidisciplinary discussion, the diagnosis was performed with ease. CT showed fat stranding in a lipomatous appendage of falciform ligament compatible with infarction (Fig.1 and 2). Although without surgical or histopathological proof, the similarities with other reported cases are striking.¹⁻³

The pathophysiology of this condition is similar to the more commonly seen omental infarction and epiploic appendagitis, encompassing the spectrum of intra-abdominal focal fat infarction (IFFI) that has recently been described.^{2,4}

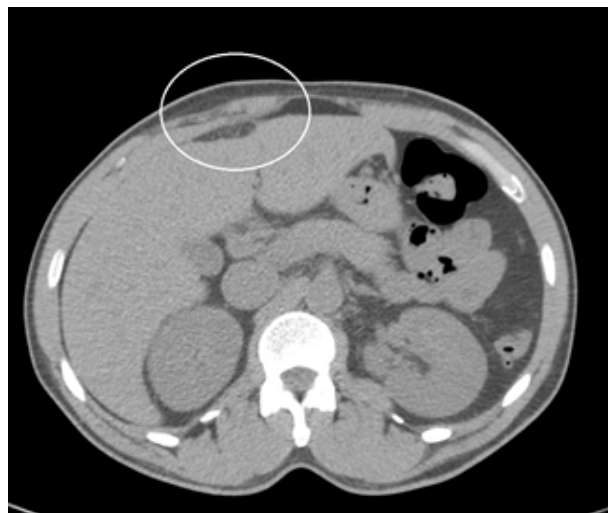


Figure 1 – Axial CT image showing fat stranding in a lipomatous appendage of the falciform ligament compatible with infarction. The surrounding inflammatory changes in the adjacent fat and myofascial planes give rise to a subtle focal cutaneous lump, evident if we compare it with the contralateral side.

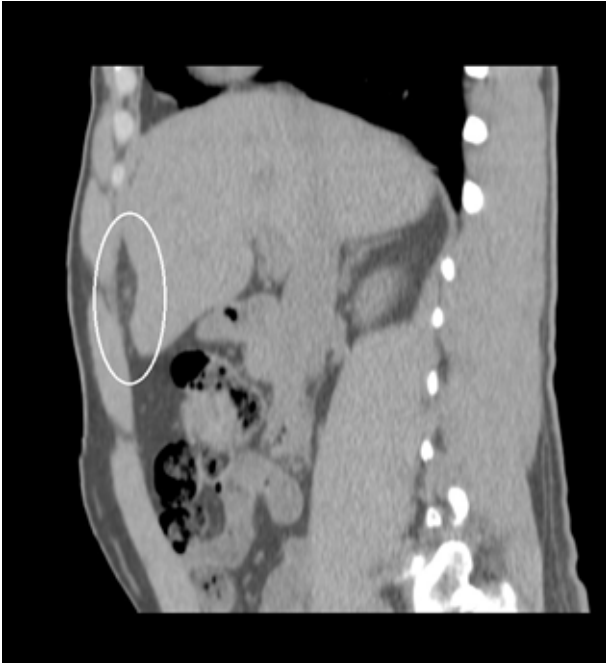


Figure 2 – Sagittal CT image depicting the longitudinal extent of the lipomatous appendage and the mass effect on liver surface with a prominent inward bulging. Also in better detail the extent of the fat stranding, and the “dot sign”, as similarly described by Uyttenhove F et al. in their case report³, caused by thrombosis of the vessels in the center of the lipomatous appendage.

Received / Recebido 13/03/2017

Acceptance / Aceite 25/05/2017

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Swienton D, Shah V. Infarction of a fatty appendage of the falciform ligament - a case report. EuroRad Online 11 April 2013. Available from: <http://www.eurorad.org/case.php?id=10799>.
2. Maccallum C, Eaton S, Chubb D, Franzi S. Torsion of fatty appendage of falciform ligament: Acute abdomen in a child. Case Rep Radiol. 2015;293491
3. Uyttenhove F, Leroy C, Nzamushe L, Lapan M, Mabl JR, Ernst O. Torsion of a fatty fringe of the falciform ligament, a rare cause of right hypochondrial pain. Diagn Interv Imaging. 2013;94:637-9.
4. Van Breda Vriesman AC, Lohle PN, Coerkamp EG, Puylaert JB. Infarction of omentum and epiploic appendage: diagnosis, epidemiology and natural history. Eur Radiol 1999;9:1886-92.

ARP Case Report n° 13: What is your diagnosis?

Caso Clínico ARP n°13: Qual o seu diagnóstico?

Ana Catarina Silva

Serviço de Radiologia, Hospital Pedro Hispano, Matosinhos, Portugal

Presentation of the Case

A 73-year-old woman who has been consulting the gynaecologist regarding persistent pelvic pain since last month. No other complaints including systemic symptoms or metrorrhagia.

The ultrasound study showed a large pelvic tumour mass and it was not possible to determine the organ of origin. Values of Ca-125 are normal.

Pelvic MRI was requested for additional characterization of the pelvic mass and to determine the organ of origin.

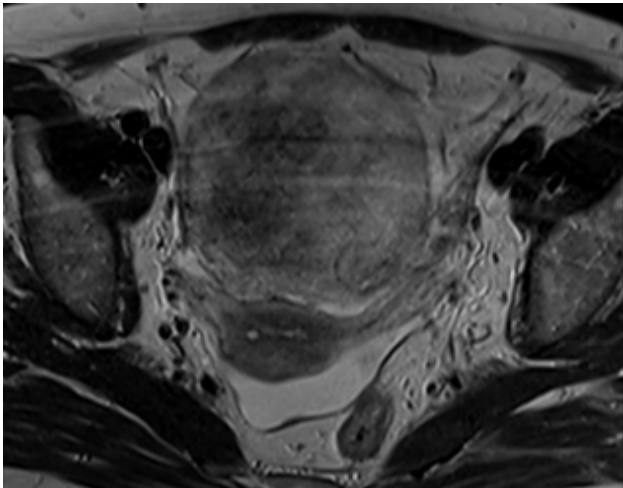


Figure 1 – Axial T2

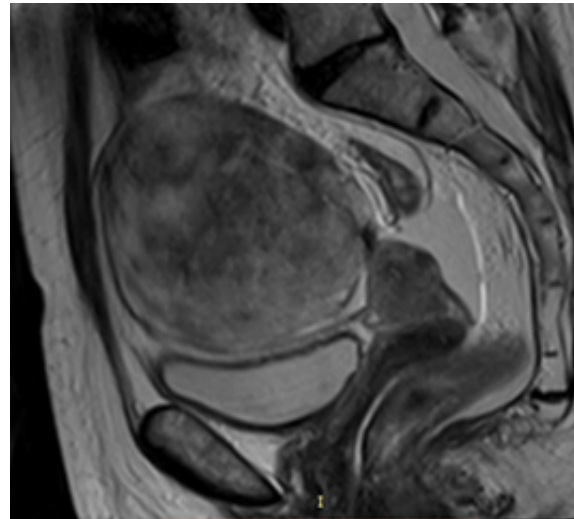


Figure 2 – Sagittal T2

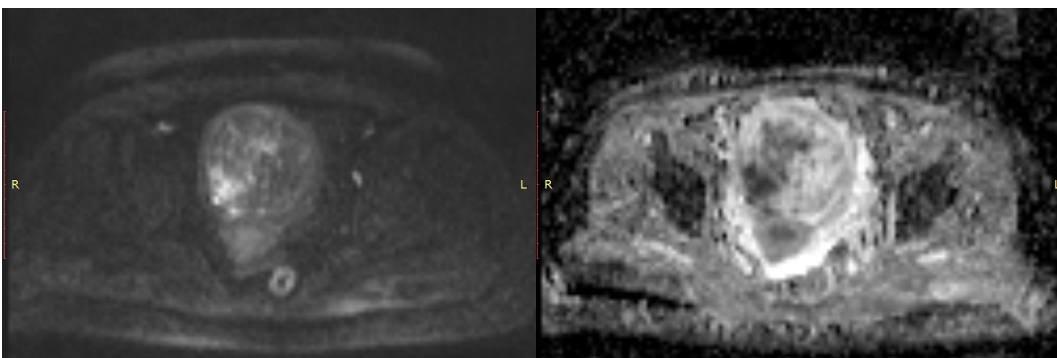


Figure 3 – Diffusion (b 1000) with corresponding ADC

Send your answer containing the diagnosis (s) to the email address actarp.on@gmail.com.
The names of the authors of the correct answers will be published in the next issue of the ARP in the case solution.

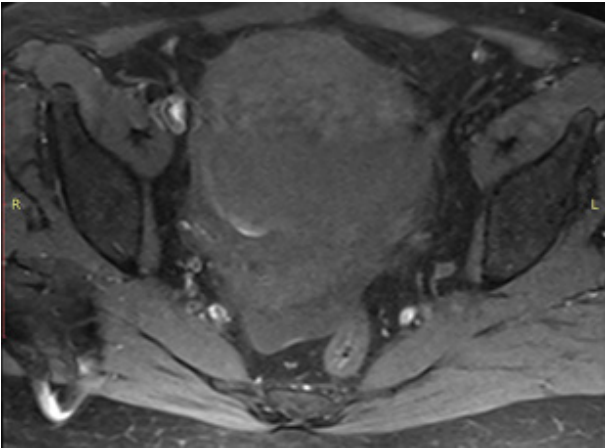


Figure 4 – Axial T1 with fat saturation

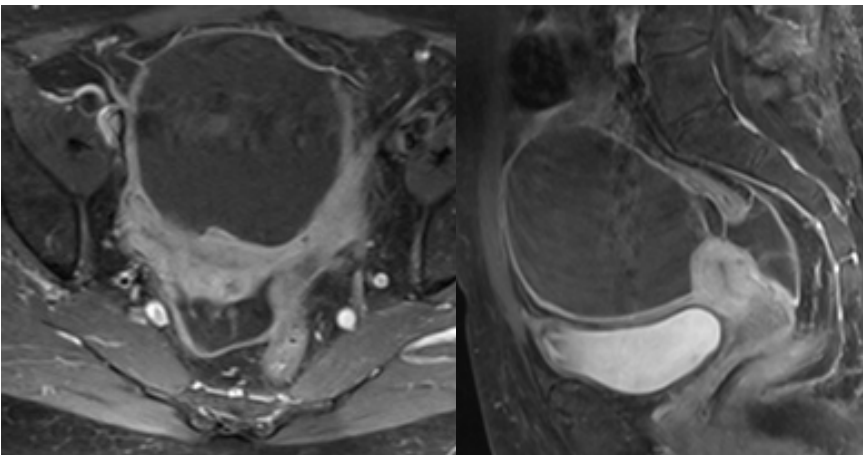


Figure 5 – Axial and sagittal T1 with fat saturation after administration of EV contrast

ARP Case Report N° 12: Communicating Varix between the Left Renal Vein and Left Ascending Lumbar Vein

Caso Clínico ARP N°12: Variz em Comunicação entre Veia Renal Esquerda e a Veia Lombar Ascendente Esquerda

Carlos Francisco Silva

Imaging Department, Setubal Hospital Centre,
Portugal

Address

Carlos Francisco Silva
Serviço de Imagiologia
Centro Hospitalar de Setúbal
Setúbal, Portugal
e-mail: carlos.f.silva@chs.min-saude.pt

Correct Answers Clinical Case N° 12

Alcinda Reis
Nuno Costa

Abstract

69-year-old female found on a routine radiological follow-up (yearly abdominal CT scan) after a left adrenalectomy 4 years ago (pathologically proven cortical adenoma). Chronic left flank pain, already present before the adrenalectomy was the major complaint.

A small left para-aortic “mass” was the main finding on the CT scan. As differential diagnosis we have considered para-aortic lymphadenopathy, adrenal mass or a saccular renal artery aneurysm given a somewhat prominent contrast enhancement.

This patient had previous abdominal CT scans, one before the adrenalectomy and the others after that surgery, and in all but one of them it was retrospectively shown that this “mass” was already present.

Coronal and oblique axial views and MIP reconstruction better depicted that the “mass” was indeed a varix or varicosity that put into communication the left renal vein and the left ascending lumbar vein.

Literature review have shown that this varix could be an explanation for the chronic left pain that this patient had because of the compression and irritation of the left lumbar plexus given the close relation of these two anatomical structures. The “disappearance” or transitory collapse of this varix in one of the patient’s previous follow-up CT scans that we retrospectively analyzed might be due to variations in intra-abdominal pressure related to Valsalva maneuver during the CT scan image acquisition.

After alerting the requesting physicians to this varix and possible explanation for the chronic pain complaints, the patient was referred to Pain Medicine specialty in our institution.

Resumo

Mulher de 69 anos de idade em seguimento radiológico de rotina - tomografia computadorizada (TC) abdominal anual - após uma adrenalectomia esquerda há 4 anos (adenoma cortical comprovado patologicamente). Queixa principal de dor crónica no flanco esquerdo, já presente antes da adrenalectomia.

Uma pequena massa para-aórtica esquerda foi a principal descoberta na TC. Como diagnóstico diferencial, consideramos a linfadenopatia para-aórtica, a massa adrenal ou um aneurisma sacular da artéria renal dada uma captação de contraste um tanto ou quanto proeminente.

Estavam disponíveis TCs abdominais prévias, uma antes da adrenalectomia e as demais após a cirurgia, e em todas, exceto uma delas, foi demonstrado retrospectivamente que essa “massa” já estava presente.

Nos cortes coronal e axial oblíquo bem como na reconstrução MIP foi melhor demonstrado que a “massa” era de facto uma variz ou varicosidade que punha em comunicação a veia renal esquerda e a veia lombar ascendente esquerda.

A revisão da literatura mostrou que esta variz poderia ser uma explicação para a dor crónica do flanco esquerdo que esta paciente apresentava devido à compressão e irritação do plexo lombar esquerdo, dada a íntima relação destas duas estruturas anatómicas.

O “desaparecimento” ou o colapso transitório dessa variz numa das TCs da paciente que analisamos retrospectivamente pode ser devido a variações na pressão intra-abdominal relacionadas com a manobra de Valsalva durante a aquisição das imagens na TC.

Após terem sido alertados os médicos assistentes sobre essa variz e possível explicação para as queixas de dor crónica, a paciente foi encaminhada para a especialidade de Medicina da Dor no nosso centro hospitalar.

Clinical Case

A 69-year-old female was found on a routine radiological follow-up (yearly abdominal CT scan) after a left adrenalectomy 4 years ago (pathologically proven cortical adenoma, after removal, documented on our institutional electronic medical records). She complained of chronic left flank pain, already present before the adrenalectomy. She also had prior surgical history of intestinal volvulus 18 years ago. This patient had previous abdominal CT scans in our institutional digital imaging archive (PACS), one before the left adrenalectomy and the others after that surgery, and in all

but one of them it was retrospectively shown that a small left para-aortic “mass” was already present (Figure 1).

As differential diagnosis of this para-aortic “mass” we could consider para-aortic lymphadenopathy, adrenal mass or a saccular renal artery aneurysm given a somewhat prominent contrast enhancement.

Coronal view and MIP reconstruction (Figures 2 and 3, respectively) show that the “mass” is indeed a varix or varicosity communicating with the left renal vein. Axial and oblique axial images (Figures 4 and 5, respectively) more clearly depict the ascending lumbar veins and the communication of the varix with the left ascending lumbar vein.

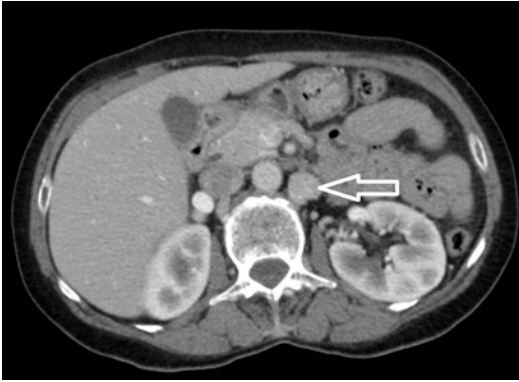


Figure 1 - Axial CT image demonstrating a small left para-aortic "mass" (arrow).



Figure 2 - Coronal view showing that the "mass" (arrow) is indeed a varix or varicosity communicating with the left renal vein.

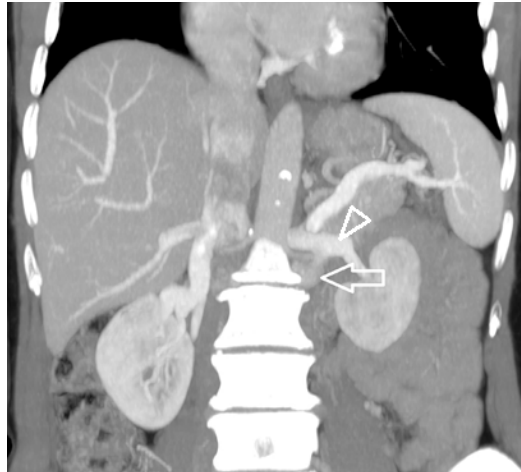


Figure 3 - MIP reconstruction putting into better detail the relationship of the varix (arrow) and the left renal vein (arrowhead).

The similarities of this case with other case reports are striking, namely with that from Jakhere et al,¹ where they state that this entity is rare with only a few cases reports in the literature, and that this could be an explanation for the chronic left pain that this patient had because of the compression and irritation of the left lumbar plexus given the close relation of these two anatomical structures.

So, interestingly in one CT scan, nearly in the middle of this 4 year period of follow-up, the varix was not evident. It could be that the "disappearance" or transitory collapse of this varix in one of the patient's previous follow-up CT scans that we retrospectively analyzed might be due to variations in intra-abdominal pressure related to Valsalva

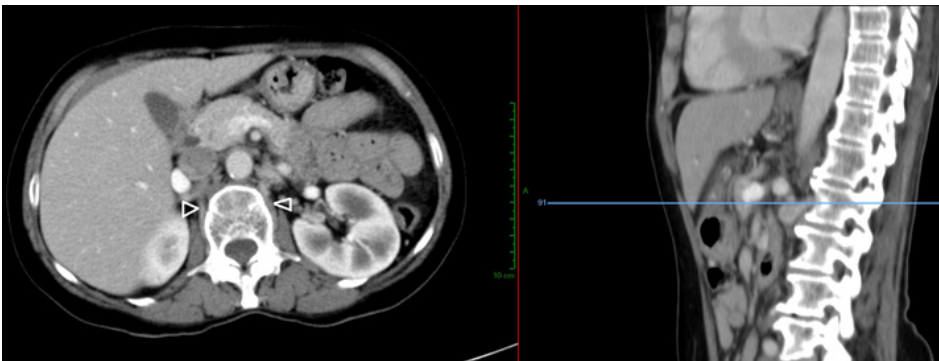


Figure 4 - Axial CT image, and the corresponding level on sagittal at the right side of the image, showing the right and left ascending lumbar veins (arrowheads); the left vein is somewhat more prominent than the right one.

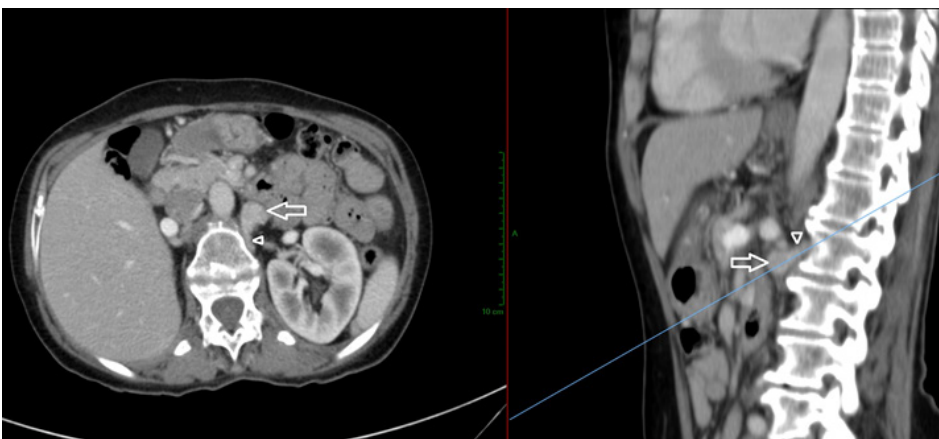


Figure 5 - Axial oblique CT image, and the corresponding level on sagittal at the right side of the image, putting in to better detail the communication of the varix (arrow) with the left ascending lumbar vein (arrowhead).

maneuver during the usual deep inspiration and apnea requested for CT scans.

After alerting the requesting physicians to this varix and possible explanation for the pain complaints, the patient was referred to Pain Medicine specialty in our institution.

References

1 - Jakhere SG, Yadav DA, Tuplondhe GR. Case report: Varicosity of the communicating vein between the left renal vein and the left ascending lumbar vein mimicking a renal artery aneurysm: report of an unusual site of varicose vein and a novel hypothesis to explain its association with abdominal pain. *Indian J Radiol Imaging*. 2011;21:24-7.

Radiological Case Report / Caso Clínico

Duodenal Hematoma after upper Gastrointestinal Endoscopy: Case Report and Literature Review*Hematoma Duodenal Pós Endoscopia Digestiva Alta em Idade Pediátrica: Caso Clínico e Revisão da Literatura***Marta Reis de Sousa, Ana Catarina Vieira, Gisela Rio, Ângela Moreira, Maria José Noruegas, Conceição Sanches**Centro Hospitalar e Universitário de Coimbra –
Hospital Pediátrico, Coimbra, Portugal**Address**Marta Reis de Sousa
Rua Nova de S. Crispim, 244, 3o
4000-363 Porto, Portugal
email: martareisdesousa@gmail.com**Abstract**

Duodenal hematoma is a rare complication of endoscopic duodenal biopsy, with just a few cases reported in children in the literature available.

The authors present a case of a 13 year-old girl, with a history of Noonan Syndrome and neurofibromatosis type 1, who presented abdominal pain and vomiting after an endoscopic duodenal biopsy.

In this article, we describe the clinical case, imaging findings, evolution and therapeutic approach.

We briefly discuss the hematologic complications in patients with Noonan syndrome.

A review of the literature and data from similar cases reported are briefly presented and discussed.

Keywords

Digestive system abnormalities; Duodenal hematoma; Endoscopy.

Resumo

O hematoma duodenal é uma complicação rara da endoscopia digestiva alta com biópsia duodenal, com poucos casos em idade pediátrica descritos na literatura.

Os autores descrevem o caso de uma adolescente de 13 anos, com antecedentes de síndrome de Noonan, neurofibromatose tipo I, que após endoscopia digestiva alta inicia quadro de dor abdominal e vômitos.

Neste artigo são descritos a apresentação clínica, os achados imagiológicos, bem como a evolução e a terapêutica proposta.

É feita uma breve apresentação e discussão de casos descritos na literatura.

É feita uma breve revisão acerca das alterações hematológicas em pacientes com síndrome de Noonan.

Palavras-chave

Anomalias do sistema digestivo; Hematoma duodenal; Endoscopia.

Clinical Case

A 13-year-old female, with a history of Noonan syndrome, neurofibromatosis type I, growth retardation, weight between P10 and P25, and stature below P5, followed in multiple specialties at our institution.

No regular medication.

Due to the presence of recurrent episodes of epigastric abdominal pain, an upper endoscopy (UE) was performed which revealed erythematous gastritis. Antral and duodenal biopsies were performed. The endoscopy proceeded without immediate complications, with normal progression of the endoscope.

24 hours after endoscopy, the patient presents with vomiting, nausea and persistent epigastralgia with pyrosis. She was hemodynamically stable.

Analytically, there was elevation of pancreatic enzymes with amylase 252 U / L (normal 30-110) and lipase 542 U / L (normal 23-300). The remaining analytical parameters were normal, namely hemoglobin, platelet count and coagulation tests (INR, TP, aPTT).

Abdominal radiography revealed no significant changes (Fig. 1). An abdominal ultrasound was performed, which revealed a heterogeneous collection anterior to the pancreas, and free intraperitoneal fluid in the pelvis. (Fig 2).

Subsequently, a computed tomography was performed for better characterization, which revealed findings compatible with intramural duodenal hematoma (Fig. 3).

She was submitted to conservative treatment with symptomatic improvement: pain control, intravenous hydration, nutritional support and antibiotics (ampicillin, gentamicin and metronidazole). She presented a good clinical and analytical evolution.

Imaging wise ultrasound control with progressive reduction of the dimensions of the described collection was performed.

Discussion

Intramural duodenal hematoma is a rare pathology in pediatric patients, occurring more frequently after blunt abdominal trauma.^{1,2} The incidence of intramural duodenal hematoma as a complication of UE with duodenal biopsy is not known, with 18 cases described in the pediatric literature.¹ It is estimated to occur in 1 in every 1250 UEs.¹ Among the known risk factors for its occurrence are coagulation and hemostasis disorders, malnutrition, and growth retardation.^{3,4,5} However, in 28 cases described in the literature, both in adults and children, only 6 had changes in the coagulation tests or platelet dysfunction.¹

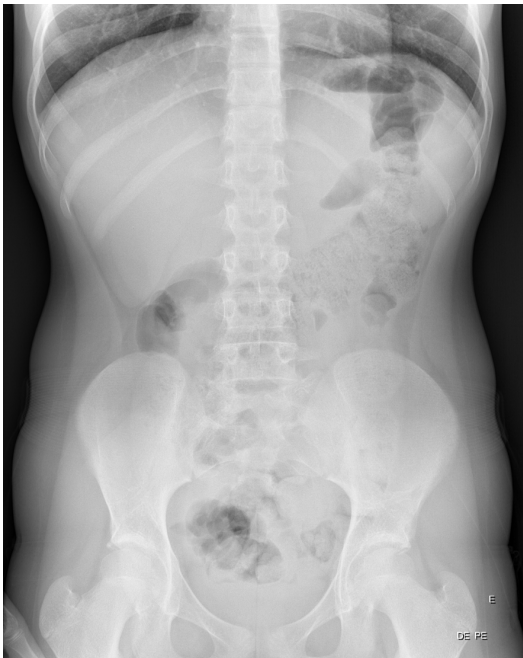


Figure 1 – Abdominal radiograph, standing, without significant changes, namely signs of pneumoperitoneum.

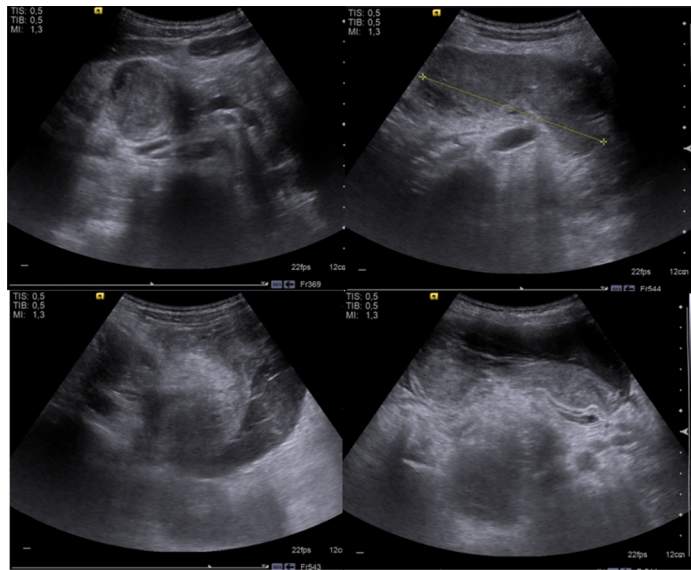


Figure 2 – Abdominal ultrasound reveals elongated collection, measuring 12 cm x 5 cm (LxAP) with heterogeneous echogenic content and fluid/fluid level, located anteriorly to the pancreas.

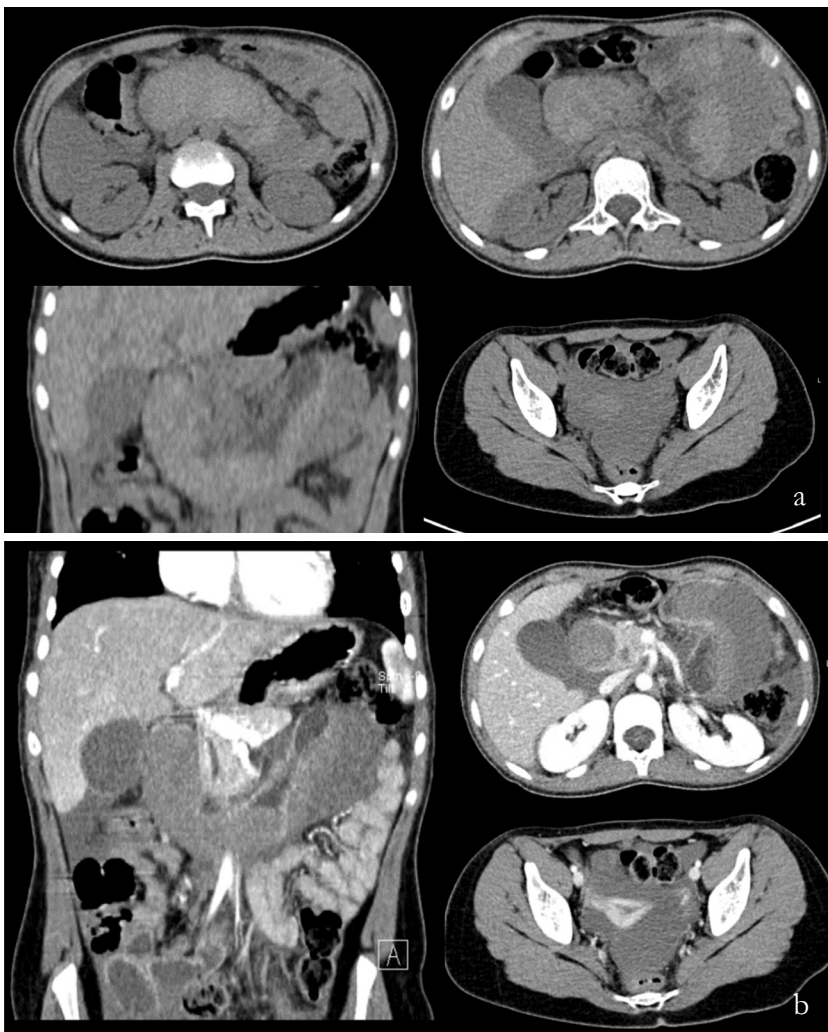


Figure 3 – Computed tomography before (a) and after (b) intravenous contrast, with axial and coronal reconstructions reveals a heterogeneous collection, spontaneously hyperdense, without contrast enhancement, that follows the duodenum from the second to fourth portion, compatible with intramural duodenal hematoma. Absence of dilation of the bile ducts. The pancreas presents normal dimensions and homogenous enhancement. Moderate amount of fluid in the pelvic excavation.

This complication has also been described in patients with leukemia or a history of bone marrow transplantation.³ Intramural duodenal hematoma is frequently associated with acute pancreatitis and is probably related to the presence of ampullary hematoma with obstruction of the papilla, or to the compression exerted on the pancreas by the hematoma.^{6,7} There are two cases described in the literature of patients with Noonan syndrome who developed duodenal hematoma as a complication of UE with duodenal biopsy. The authors associate this complication with the frequent presence of growth retardation and coagulation and hemostasis alterations in these patients.^{5,7} Hemorrhagic problems have been reported in approximately 55% of these patients.⁶ Noonan syndrome may be associated with deficiencies of coagulation factors (factor VIII, XI, XII), thrombocytopenia and platelet dysfunction.^{8,9} In these patients coagulation tests with platelet count, aPTT, TP is recommended. In some cases platelet function tests and clotting factor counts may be required.⁹ In the clinical presentation, symptoms caused by duodenal obstruction, with nausea and vomiting, predominate. If symptoms of abdominal pain and vomiting occur in the first 48 hours after duodenal biopsy, the diagnostic hypothesis of intramural hematoma should be considered.¹ The diagnosis is confirmed by imaging, followed by conservative treatment, usually with good prognosis, with an estimated resolution time of two to three weeks.^{2,1} Surgical approach may be indicated if there is suspicion or confirmation of perforation or in the absence of improvement with conservative treatment.

Abdominal ultrasound is a rapid and accessible examination that allows evaluation of the presence of duodenal hematoma as well as its evolution.

Computed tomography allows a better characterization and evaluation of the hematoma extent, allows the diagnosis of perforation, which is why it should be performed early.¹ MRI also allows characterization of lesions and evolutionary control.

Upper gastrointestinal series may be useful to demonstrate duodenal obstruction,¹ evidencing an obstructive mass or diffuse thickening of the duodenal folds,² but it may underestimate the lesion extent.

Ultrasound guided drainage may be considered if there is no reduction in size of the hematoma within in 7-14 days.¹

Conclusion

Intramural duodenal hematoma is a rare complication after UE with biopsy. The diagnosis is confirmed by imaging. This complication must be known and considered by radiologists when after UE with duodenal biopsy the patient presents with nausea, vomiting and abdominal pain, since early diagnosis, management and evaluation of associated complications is crucial. Ultrasonography is the first-line examination, and computed tomography allows better characterization, evaluation of lesion extent and detection of other associated complications.

Received / Recebido 13/03/2017

Acceptance / Aceite 25/05/2017

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Grasshof C, Wolf A, Neuwirth F, Posovszky C. Intramural duodenal haematoma after endoscopic biopsy: case report and review of the literature. *Case Reports in Gastroenterology*. 2012;6:5-14.

2. Borsaru AD, Nandurkar D. Intramural duodenal haematoma presenting as a complication after endoscopic biopsy. *Australas Radiol*. 2007;51:378-80.

3. Diniz-Santos DR, de Andrade Cairo RC, Braga H, Araújo-Neto C, Paes IB, Silva LR. Duodenal hematoma following endoscopic duodenal biopsy: a case report and review of the literature. *Can J of Gastroenterol*. 2006;20:39-42.

4. Ghishan FK, Werner M, Vieira P, Kuttles J, DeHaro R. Intramural duodenal hematoma: an unusual complication of endoscopic small bowel biopsy. *Am J Gastroenterol*. 1987;82:368-70.

5. Sgouros SN, Karamanolis G, Papadopoulou E, Papageorgiou G, Stefanides G, Nastos H, Mantides A. Postbiopsy intramural hematoma of the duodenum in an adult with noonan's syndrome. *J Gastroenterol Hepatol*. 2004;19:1217-19.

6. Leva E, Macchini F, Cesare A, Arnoldi R, Gentilino V. Duodenal hematoma and pancreatitis complicating endoscopic intestinal biopsy in a boy with noonan syndrome. *J Trauma Treatment*. 2012.

7. Shiozawa K, Watanabe M, Igarashi Y, Matsukiyo Y, Matsui T, Sumino Y. Acute pancreatitis secondary to intramural duodenal hematoma: case report and literature review. *World J Radiol*. 2010;2:283-88.

8. Burgt Ineke van der. Noonan syndrome review. *Orphanet Journal of Rare Diseases*. 2007;2:4.

9. Briggs B, Dickerman J. Bleeding disorders in noonan syndrome. *Pediatric Blood and Cancer*. 2012;58:167-72.

Radiological Case Report / Caso Clínico

Intraarticular Osteoid Osteoma of the Elbow – A Challenging Case*Osteoma Osteóide Intra-Articular no Cotovelo – Um Desafio Diagnóstico*Natália Ferreira¹, António Costa², Ana Ferreira¹, Artur Duarte³, José Fonseca Santos⁴

¹Interna complementar de Radiologia do Serviço de Imagiologia Geral do Centro Hospitalar Lisboa Norte, Lisboa, Portugal

²Assistente hospitalar do Serviço de Radiologia do Hospital de Vila Franca de Xira, Vila Franca de Xira, Portugal

³Assistente hospitalar do Serviço de Imagiologia Geral do Centro Hospitalar Lisboa Norte, Lisboa, Portugal

⁴Diretor do Serviço de Imagiologia Geral do Centro Hospitalar Lisboa Norte, Lisboa, Portugal

Address

Natália Ferreira
Passeio do Adamastor
lote 5, 2º esq.
1990-007 Lisboa, Portugal
email: nataliasanferreira@hotmail.com

Abstract

Osteoid osteoma is a common bone tumor, usually found in young patients. Intraarticular locations are rare, occurring in approximately 13% of cases. The most commonly involved joint is the hip, while the elbow is less commonly affected.

Intraarticular osteoid osteoma may be associated with atypical clinical features and imaging findings that often differ from the classical hallmarks of extraarticular lesions.

Patients with osteoid osteoma of the elbow frequently present pain, chronic synovitis, joint effusion and limitations in motion, simulating inflammatory arthropathy. Additionally, in intraarticular lesions, reactive cortical thickening or sclerosis is minimal or absent giving a subtle radiographic appearance that often delays the diagnosis. Careful search for history data and extensive imaging procedures with computed tomography, bone scintigraphy and magnetic resonance can lead to the correct diagnosis.

The case of a young male with an osteoid osteoma of the elbow is presented.

Keywords

Osteoid osteoma; Bone neoplasms; Diagnostic imaging; Tomography; Magnetic resonance imaging

Resumo

O osteoma osteóide é um tumor ósseo comum que ocorre predominantemente em jovens. A localização intra-articular é rara, surgindo em cerca de 13% dos casos. A anca é a articulação mais vezes envolvida, enquanto que o cotovelo é uma localização menos comum.

O osteoma osteóide intra-articular pode apresentar-se com manifestações clínicas atípicas e achados imagiológicos que diferem dos sinais clássicos das lesões extra-articulares.

Os casos de osteoma osteóide do cotovelo frequentemente cursam com dor, sinovite crónica, derrame articular e limitação de movimento, simulando uma artropatia inflamatória. Acresce que nas lesões intra-articulares o espessamento cortical ou esclerose podem ser mínimos ou ausentes, dificultando a deteção das lesões pela radiografia. A avaliação detalhada da história clínica e o recurso a métodos de imagem como a tomografia computadorizada, cintigrafia óssea e ressonância magnética são essenciais na abordagem diagnóstica.

Os autores apresentam um caso de osteoma osteóide do cotovelo num jovem do sexo masculino.

Palavras-chave

Osteoma osteóide; Tumores ósseos; Diagnóstico radiológico; Tomografia; Ressonância magnética.

Introduction

Osteoid osteoma is a relatively common bone lesion and corresponds to approximately 10-12% of all benign bone tumors.¹ It is usually found in young individuals between the ages of 7 and 25 years and men are more frequently affected than women.^{1,2} Patients often show pain that worsens at night and is relieved by the administration of salicylates.¹ Osteoid osteoma is preferentially found in the diaphysis of the femur and the tibia.³

Intraarticular osteoid osteoma, which occurs within or near a joint, is rare and considered a separate clinical entity.^{2,4} Intraarticular locations may be associated with atypical clinical features and imaging findings often differ from those of intracortical osteoid osteoma.² When intraarticular, reactive cortical thickening or sclerosis is minimal or absent giving a subtle radiographic appearance that often leads to delay of diagnosis.⁵

Computed tomography (CT) is the method of choice to identify the nidus, whereas magnetic resonance imaging (MRI) is the best technique to identify changes in the intramedullary area and in the soft tissues adjacent to the lesion.⁶

We present a case of a boy with an intraarticular osteoid osteoma of the elbow.

Clinical History

A 15-year-old male presented progressive, mild intensity pain and stiffness of the elbow for a 3-month period. There was no history of trauma and his past medical history was unremarkable. On physical examination, the patient reported pain on elbow motion. The plain film did not reveal significant findings.

An ultrasonography of the elbow was performed, revealing hypoechoic synovitis with associated hyperemia on the

color Doppler evaluation and a small joint effusion (fig. 1). Intense pain was produced when the transducer was pressed over the area of the synovitis.

The patient underwent a contrast-enhanced MR imaging examination. A small, round cortical lesion with intermediate signal intensity on T1- and T2-weighted MR images and modest enhancement was noticed in the coronoid fossa of the distal extremity of the elbow (fig. 2). Bone marrow edema adjacent to the lesion, synovitis and mild joint effusion were also present (fig. 2). The diagnosis of osteoid osteoma was suspected.

Finally, a CT was performed, revealing a 15-mm lytic lesion with a central radiolucent nidus that contained a calcified center, located in the coronoid fossa of the distal extremity of the elbow, bulging through the cortical bone (fig. 3). Periosteal reaction and subtle cortical thickening was noted. The CT findings confirmed the diagnosis of intraarticular osteoid osteoma.

After the lesion was surgically removed, the patient remained asymptomatic.

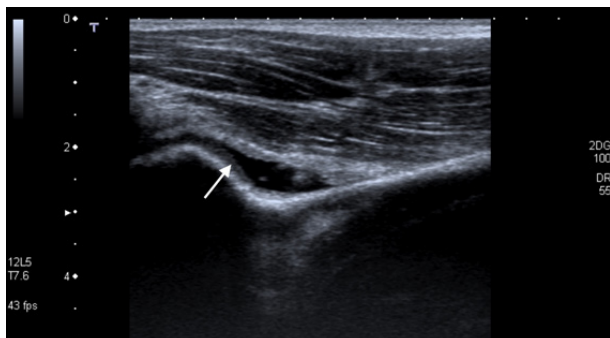


Figure 1 – Ultrasound of the elbow depicting synovitis (arrow) with mild joint effusion.



Figure 2 – MRI of the elbow. Sagittal images on T1 TSE (a) and PD TSE SPAIR (b) show the nidus with isointense signal on T1 and slightly hyperintense signal on proton density weighted images (arrow). Note periosteal reaction (curved arrows) and synovitis with mild joint effusion (*). Axial T1 SPIR after gadolinium administration (c) depicts slight enhancement of the nidus (arrow). Adjacent bone marrow edema is also observed.

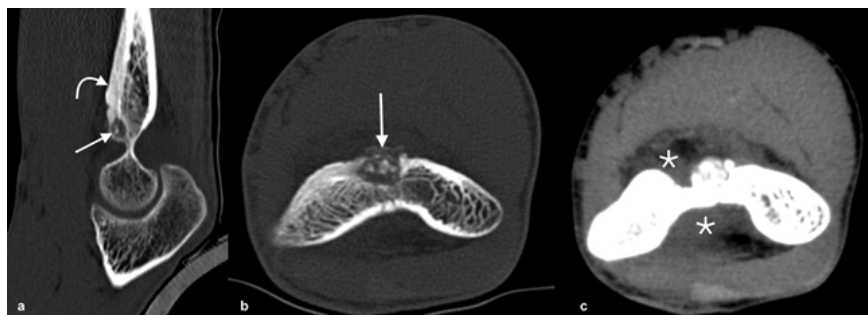


Figure 3 – CT of the elbow. Sagittal reconstruction (a) demonstrates the presence of the radiolucent nidus in the coronoid fossa (arrow) and prominent periosteal reaction (curved arrow). Axial CT image with bone window (b) show the 15-mm diameter radiolucent nidus (arrow) that contains a calcified centre. On axial CT image with soft tissue window (c) mild joint effusion is identified.

Discussion

Approximately 13% of osteoid osteoma arise within a joint.⁷ This tumor is considered to be intraarticular when it occurs at the extremity of long bones, around or within the joint, in a bone limited by the capsule, very close to it

and to the synovia¹. The most commonly involved joint is the hip, while the ankle, elbow, wrist, and knee are less commonly affected.²

In intraarticular osteoid osteoma, clinical symptoms can be different from the classical hallmarks of extraarticular lesions. The pain is not necessarily worse at night and may not be relieved by salicylates.⁷ Additionally, joint tenderness and effusion may be prominent, contributing to the diagnostic confusion.²

Patients with osteoid osteoma of the elbow frequently present pain, chronic synovitis, joint effusion and limitation in range of motion, simulating inflammatory arthropathy,¹ as in this case. Furthermore, the radiological findings can be uncharacteristic and misleading.

At plain film and CT, osteoid osteoma is typically depicted as a well-defined, round or ovoid lytic lesion, called nidus, that is surrounded by an area of bone sclerosis and/or reactive cortical thickening.² CT is the method of choice to identify the nidus of an intra- or juxta-articular osteoid osteoma.³ In the center of the nidus a focus of high attenuation can be seen, a feature corresponding to mineralized osteoid. In cases of intraarticular involvement, reactive cortical thickening can be minimal or absent,^{1,5} reason why sometimes it is difficult to identify it on the plain film.

On MR imaging, the nidus shows low or intermediate signal on T1-weighted images and has variable signal on T2-weighted images.² High signal in the bone marrow and soft-tissue abnormalities on T2-weighted images may be found adjacent to the lesion.² Compared with CT, MRI is less sensitive to depict small nidi because the signal is similar to that in cortical bone¹. Enhancement of the nidus may be seen both in CT and MRI after contrast intravenous administration.

On bone scintigraphy, the tumour presents a typical pattern known as “double density” sign: intense activity centrally in the nidus region and less intense activity in the periphery of the lesion.³ However, bone scintigraphy often fails to visualize the nidus and shows unspecific findings.⁶

The differential diagnosis for osteoid osteoma of the elbow include: monoarthritis, osteochondritis dissecans, osteomyelitis, chondroblastoma and osteoblastoma. Complete nidus excision is curative and is the most traditional treatment method. However, minimally invasive techniques, such as radiofrequency ablation, have also been proved safe and effective alternatives.³ There's also evidence that osteoid osteoma may resolve spontaneously over time

Received / Recebido 10/09/2017

Acceptance / Aceite 23/12/2017

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

and can be treated conservatively with nonsteroidal anti-inflammatory drugs in certain patients.³

In conclusion, clinicians and radiologists should be aware of the potentially confusing clinical and imaging findings associated with intra-articular osteoid osteoma. Careful search for history data and extensive imaging procedures can lead to the correct diagnosis.

References

1. Cotta AC, Melo RT, Castro RC, Souza FS, Najjar YS, Paim JF, et al. Diagnostic difficulties in osteoid osteoma of the elbow: clinical, radiological and histopathological study. *Radiol Bras.* 2012;45:13–9.
2. Chai JW, Hong SH, Choi JY, Koh YH, Lee JW, Choi JA, et al. Radiologic diagnosis of osteoid osteoma: from simple to challenging findings. *Radiographics.* 2010;30:737–49.
3. Traore SY, Dumitriu DI, Docquier PL. Intra-articular osteoid osteoma mimicking juvenile arthritis. *Case Rep Orthop.* 2014;912609.
4. Kattapuram SV, Kushner DC, Phillips WC, Rosenthal DI. Osteoid osteoma: an unusual cause of articular pain. *Radiology.* 1983;147:383–7.
5. Ebrahim FS, Jacobson JA, Lin J, Housner JA, Hayes CW, Resnick D. Intraarticular osteoid osteoma: sonographic findings in three patients with radiographic, CT, and MR imaging correlation. *Am J Roentgenol.* 2001; 77:1391–5.
6. Szendroi M, Köllö K, Antal I, Lakatos J, Szoke G. Intraarticular osteoid osteoma: clinical features, imaging results, and comparison with extraarticular localization. *J Rheumatol.* 2004;31:957–64.
7. Allen SD, Saifuddin A. Imaging of intra-articular osteoid osteoma. *Clin Radiol.* 2003;58:845–52.

Radiological Case Report / Caso Clínico

Spontaneous Hepatic Haemorrhage of Unknown Cause – A Case Report

Hemorragia Hepática Espontânea de Causa Desconhecida - Um Caso Clínico

Elizabeth Cabral Matos

Centro Hospitalar Vila Nova de Gaia, Espinho,
Portugal

Address

Elizabeth Cabral Matos
Centro Hospitalar de Vila Nova de Gaia
Serviço de Imagiologia
Rua Conceição Fernandes S/N
4434-502 Vila Nova de Gaia, Portugal
email: elizabeth.cabral.matos@gmail.com

Abstract

To report a clinical case of spontaneous hepatic haemorrhage with hemoperitoneum with no defined underlying cause. A detailed analysis and description of the spectrum of ultrasonographic (US), tomodesitometric (TDM) and angiographic findings - important diagnostic tools in this clinical setting – are done, in line with a review of the state of the art literature.

A 52-year old woman with a poorly defined and inconclusive past medical history of gallstones and an autoimmune disorder treated with Salazopyrin complained of acute abdominal pain and vomiting. She was hospitalized with an initial diagnosis of acute pancreatitis. On the 6th day of hospitalization, US and TDM studies were done due to progressing abdominal pain with analytic worsening and new-onset of haemodynamic instability, although haemoglobin levels were normal. Liver haemorrhage with a large subcapsular haematoma, active parenchymal bleeding foci and hemoperitoneum was found. Initial and emergent surgical management with perihaptic packing was done. Due to active bleeding and haemodynamic instability nonresponsive to medical resuscitation, right hepatic artery angiography and embolization was performed the following day. A second laparotomy due to haemodynamic unresponsiveness, with “en bloc” evacuation of the hematoma and hepatic devitalized sequestrum, and haemostasis of the identified bleeding foci was performed two days afterwards. The patient eventually died from hemorrhagic shock.

Keywords

Spontaneous hepatic haemorrhage;
Ultrasonographic findings of hepatic haemorrhage; Tomographic findings of hepatic haemorrhage; Management of spontaneous hepatic haemorrhage.

Resumo

É apresentado um caso clínico de hemorragia hepática espontânea com hemoperitoneu sem causa subjacente definida. É feita uma análise detalhada com descrição do espectro de achados ecográficos, tomodesitométricos e angiográficos – ferramentas diagnósticas importantes neste contexto clínico – em consonância com uma revisão da literatura.

Doente do sexo feminino de 52 anos - com história de litíase vesicular e antecedentes mal definidos e inconclusivos de patologia autoimune tratada com Salazopirina – que é avaliada por um quadro agudo de dor abdominal e vômitos, e hospitalizada com um diagnóstico inicial de pancreatite aguda. No 6º dia de internamento foi efetuada avaliação imagiológica com ecografia e tomografia computadorizada por dor abdominal progressiva com agravamento analítico e instabilidade hemodinâmica “de novo”, apesar dos níveis normais de hemoglobina, tendo sido demonstrados sinais de hemorragia hepática com hematoma subcapsular, focos de hemorragia parenquimatosa ativa e hemoperitoneu. Neste contexto, avançou-se para terapêutica cirúrgica emergente com “packing” perihepático, seguida, no dia seguinte, de estudo angiográfico com embolização da artéria hepática direita, por instabilidade hemodinâmica e hemorragia ativa não responsivas ao tratamento médico. Pelo menos motivo, uma segunda laparotomia foi efetuada dois dias depois, com evacuação “em bloco” do hematoma e de sequestros hepáticos devitalizados, e hemóstase dos focos hemorrágicos identificados. Apesar das medidas efetuadas, o quadro clínico culminou em morte por choque hemorrágico.

Palavras-chave

Hemorragia hepática espontânea; Achados ecográficos de hemorragia hepática; Achados tomodesitométricos de hemorragia hepática; Terapêutica da hemorragia hepática espontânea.

Introduction

Spontaneous hepatic haemorrhage (SHH), occurring in the absence of an external cause such as trauma or anticoagulant therapy, is a rare poorly understood and potentially lethal (mortality rate of up to 75% depending on the underlying cause and clinical status) surgical emergency due to liver capsular rupture with massive intra-abdominal haemorrhage,

accounting for only 1% of admissions to specialist liver units.^{1,4,5} Although more commonly associated with the rupture of an underlying hypervascular hepatic tumour, there is a wide range of other even more rare underlying conditions, such as the Haemolysis, Elevated liver enzymes, Low platelet count (HELLP) syndrome in pregnant women, coagulation disturbances such as bleeding diathesis, connective tissue diseases, infections (in developing

countries) and miscellaneous causes.^{4,5} An association with cirrhosis, without an associated primary malignancy is also found in the literature.⁶⁻¹²

Given the fact that most patients with SHH show unspecific symptoms (such as right upper quadrant or diffuse abdominal pain) and signs (malaise, nausea or hypotension), imaging – mainly dynamic multidetector computerized tomography (MDCT) plays an important role in diagnosis and management, detecting and characterizing signs of hepatic haemorrhage with or without active bleeding, extension (intra-capsular vs intraperitoneal) and possible underlying causes.^{4,5}

Therapeutic management is complex, multidisciplinary and controversial, depending on the clinical status, the source and extent of bleeding. Intraperitoneal bleeding with haemodynamically instability requires immediate surgery or selective embolization of the hepatic arteries to achieve haemostasis.^{4,13} Herein, a case-report of SHH is presented.

Clinical Case

A 52-year old female patient, with gallstones and an inconclusive past medical history of an undefined autoimmune disease (presumably, scleroderma) went to the Emergency Room with abdominal pain and vomiting that lasted for 5 days. She was under clinical investigation for skin thickening and hardening of the extremities below the knees and elbows associated with pruritus – more prominent at the inferior extremities – with plaque-type morphea round cutaneous lesions with episodes of surrounding ring erythema; with associated muscle atrophy; without evident sclerodactyly; and associated beak-shaped nose. Pulmonary function was not determined and thoracic x-ray was normal. Renal and hepatic function tests were within the normal range. A suspicion of scleroderma with diffuse and severe skin involvement was set and treatment with Salazopyrin was started one year before the current clinical situation. A cutaneous biopsy was taken, with inconclusive results. Auto-antibodies (Rheumatoid factor, Anti-nuclear antibody including Anti-Jo1 and Anti-SCL 70 and Anti-neutrophil cytoplasmic antibodies) were negative. The patient subsequently refused to maintain medical investigation. Other past history details - including recent and/or chronic viral or bacterial infections, vaccinations, malignancies, ingestion of alcohol, and chronic medication - were not included.

At initial clinical evaluation, the patient was normotensive (124/70 mmHg); with normal heart rate (87 bpm) and 97% of oxygen saturation; with a body temperature of 37°C; and had a firm and tender abdomen on the right quadrants, without signs suggesting peritoneal irritation. Analytic study revealed haemoglobin of 15 g/dL; leucocytosis of 18.4000 leucocytes with relative neutrophilia of 85,6%; bilirubinaemia of 3,10 mg/dL with predomination of direct bilirubin measuring 2,54 mg/dL; high levels of lactate dehydrogenase (LDH) of 365 U/L (1,7 x N), aspartate and alanine aminotransferases (TGO and TGP) of 432 and 568 U/L (16 x N) and pancreatic amylase and lipase enzymes of 1097 and 1241 U/L (20 x N); high levels of reactive C protein (RCP) of 2 mg/dL; normal levels of platelets around 399 x 10E3/ μ L; hypoalbuminemia (of 1,5 g/dL) and high levels of creatinine (2 mg/dL). The patient was hospitalized with the diagnosis of lithiasic acute pancreatitis – supportive care with aggressive hydration with isotonic crystalloid solution and food break was done, with

clear liquid diet starting 48 hours after. Metoclopramide was given on a SOS regime.

After six days of hospitalization, due to persisting and progressing symptoms namely abdominal pain with the new onset of peritoneal irritation signs and haemodynamic instability (hypotension of 85-80/60-55 mmHg and tachycardia of 100-120 bpm) - and worsening analytic parameters, further radiologic investigation was performed. At his point, there was normalization of pancreatic amylase levels, with: persisting leucocytosis of 15.000 leucocytes with neutrophilia and high RCP levels of 5 mg/dL; worsening hepatic parameters with 1989, 1177 and 665 U/L (9, 44 and 19 x N) of LDH, ASP and ALP levels; reduction of haemoglobin (9 g/dL), erythrocytes and haematocrit (of 27%), and a reduction of platelets (50 x 10E3/ μ L). International normalized ratio (INR) and prothrombin ratio (PR) were in the normal range of 1,2 and 73%.

Abdominal ultrasound, although limited by bowel gas, showed an enlarged liver (with a right hepatic lobe measuring 16-cm) with a diffusely altered heterogeneous parenchymal echostructure; signs of peritoneal hematic fluid seen as echogenic and heterogeneous peritoneal fluid; without other relevant signs, namely, splenomegaly, nor portal ectasia. Dynamic MDCT was performed for further evaluation, with image acquisition before and after IV contrast administration at the arterial (at 35 seconds after contrast administration), venous (70 seconds) and delayed phase (3 minutes) of enhancement, depicting:

- A large subcapsular hepatic hematoma surrounding both lobes, seen as a perihepatic heterogeneous fluid collection with areas of spontaneously high (45 UH) density from recent bleeding (solid arrow in image 2) – showing a “haematocrit effect” with fluid-fluid levels of different densities due to blood content of different ages (arrows in images 3 and 4); compressing the adjacent parenchyma, which revealed concave irregular edges and a reduced volume (best appreciated in image 4);
- Some millimetric intraparenchymal foci of active bleeding seen as foci of high density accumulation similar to adjacent vessels after contrast administration - best seen at the arterial phase - mainly in the right hepatic lobe (curved arrows in images 5 and 6);
- Ill-defined confluent parenchymal central areas of low density/low parenchymal enhancement suggesting areas of



Image 1 – Non-enhanced axial image of the pelvis. Hemoperitoneum in the Douglas pouch (arrow).

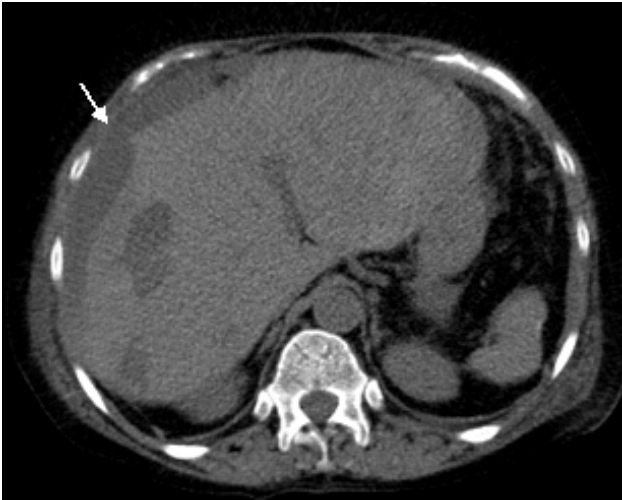


Image 2 – Non-enhanced axial image of the liver. Subcapsular liver hematoma surrounding both lobes (arrow).

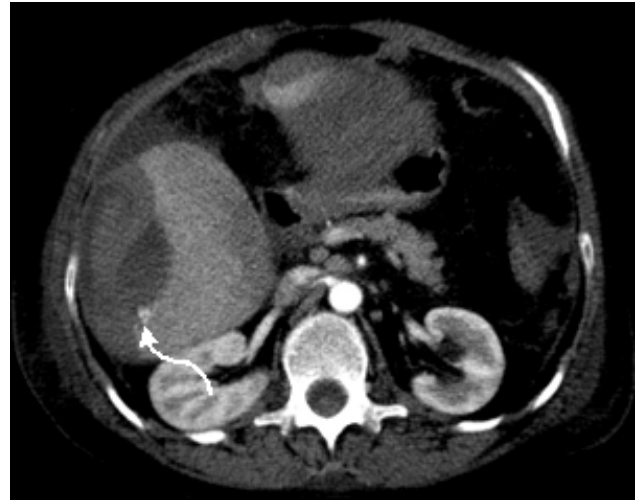


Image 5 – Enhanced axial image of the liver at the arterial phase (35 sec) of dynamic study. Subcapsular large hematoma with perihepatic intraperitoneal fluid. Millimetric foci of active parenchymal hemorrhage (curved arrow).



Image 3 – Enhanced axial image of the liver at the arterial phase (35 sec) of dynamic study. Subcapsular hepatic hematoma with fluid-fluid levels compressing the adjacent parenchyma (arrow). Confluent parenchymal areas of necrosis/ischemia (curved arrow).



Image 6 – Enhanced axial image of the liver at the portal phase (70 sec) of dynamic study. Pooling of extravasated contrast (curved arrow)



Image 4 – Enhanced axial image of the liver at the portal phase (70 sec) of dynamic study. Subcapsular hepatic hematoma with fluid-fluid levels compressing the adjacent parenchyma (arrow). Confluent parenchymal areas of necrosis/ischemia (curved arrow).

necrosis/ischemia, mainly in the left hepatic lobe (curved arrow in images 3 and 4);

- Patent filiform arteries and a normal diameter patent portal;
- A small amount of hemoperitoneum, depicted as spontaneously dense (30-40UH) content deposited in Douglas space and surrounding the liver, from capsular hepatic rupture (solid arrows in image 1 and 2; also seen in images 5 and 6).

Two units of packed red blood cells and two units of platelets were transfused, with rising of haemoglobin levels to 10 g/dL and platelets to 134 10E3/uL. One hour after US and MDCT the patient remained with tachycardia (140-150 bpm) and arterial pressures of 85-95/55-65 mmHg.

Due to hemodynamic instability, the presence of an extensive haematoma and signs of active parenchymal bleeding and hemoperitoneum, explorative laparotomy was first performed, the unique therapeutic available approach at that moment. Hemoperitoneum of large volume and hepatic haematoma with multiple capsular rupture sites were found. Peritoneal wash and perihepatic packing were done. Evacuation of subcapsular hematoma and hepatic resection were not performed in order to avoid severe haemorrhage.



Image 7 – Digital subtraction angiography of the celiac trunk with arteriography of the gastroduodenal, common hepatic artery and right and left hepatic branches. Irregularity of the right hepatic artery is seen (arrow)



Image 9 – Digital subtraction angiography after right hepatic artery embolization with coils (arrows).

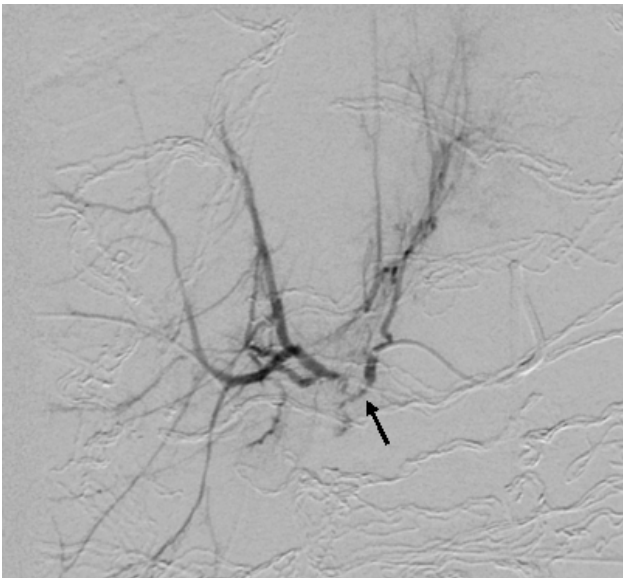


Image 8 – Digital subtraction angiography with supra-selective catheterization of the right hepatic artery better denoting the irregularity of the right hepatic artery with contrast extravasation (arrow).

Arterial embolization was unavailable in the Emergency Room at that moment.

The morning after (12 hours after surgery), hepatic artery embolization was attempted. Selective catheterization of the celiac trunk (image 7) and supra-selective catheterization of the right and left hepatic arteries (image 8) with a Progreat 2,7F catheter revealed focal irregularity in the proximal portion of the right hepatic artery (black arrow in images 7-8), with minimal contrast extravasation. Due to these findings and to limited therapeutic available options, right hepatic artery embolization was performed proximally with three microcoils of 0,018 inch. The post-embolization arteriogram (image 9) showed successful and complete embolization of this branch (white arrow). The procedure was performed without immediate complications.

Due to persistent haemodynamic instability, a second laparotomy was done (36 hours after the first surgical

approach). Direct liver exploration was done after peritoneal wash, with signs of active bleeding – through the right anterior sectorial pedicle presumably from a portal branch source. “En bloc” evacuation of the hematoma and hepatic devitalized sequestrum and haemostasis of the haemorrhagic identified focus – using a suture and Surgicel stopper - were performed. Other multiple superficial haemorrhagic foci were controlled with cauterization. After the placement of Tachosil plates and new perihepatic packing, apparent haemorrhagic control was achieved.

The patient subsequently deteriorated during the following day and died from uncontrolled bleeding.

Discussion

Spontaneous hepatic haemorrhage (SHH) is a surgical emergency with an unclear and probable multifactorial pathogenesis, related to a wide range of conditions that share a common feature of parenchymal and vasculature integrity impairment.¹ An underlying hypervascular liver tumour (such as hepatocellular carcinoma, adenoma, metastases, haemangioma, focal nodular hyperplasia and other less frequent tumours such as angiomyolipoma and angiosarcoma) is the most frequent cause.^{4,5,10} Other less frequent reported miscellaneous causes are coagulation disturbances such as bleeding diathesis (as seen in hepatic failure or thrombocytopenia) – that may precipitate or maintain bleeding;¹ vascular lesions (i.e. peliosis hepatis); inflammatory/infectious processes with microaneurysm formation; nodular regenerative hyperplasia; amyloidosis; connective tissue disorders/autoimmune disorders (best described in patients with systemic lupus erythematosus, polyarteritis nodosa or myositis); and cirrhosis without an associated with an underlying hepatocellular carcinoma - from macronodular cirrhosis and venous/lymphatic ectasias; with few case reports in the literature.^{1,4,5,12} SHH with rupture is also a known complication of some cases of pre-eclampsia and eclampsia in pregnancy, associated with the Haemolysis, Elevated liver enzymes, Low platelet count (HELLP) syndrome.⁴

In the setting of autoimmune disorders, it is believed that a possible underlying pathologic mechanism is an impaired integrity with a weakened tissue that is vulnerable to a trivial or physiological event.¹ Additionally, connective tissue or autoimmune disorders can be associated - concomitantly or serially - with a previous liver dysfunction, that can appear as a manifestation of the underlying disease or can reflect a primary liver disease with a common immunological pathogenic background.^{6,7,11} Diagnosis is made by serological, clinical and finally histological characteristics after exclusion of other potential causes.⁷

The acute thrombocytopenia found in this case was hypothesized to be from accelerated destruction and/or multifactorial, presumably with an immune background - immune thrombocytopenia (ITP) and/or drug-induced immune thrombocytopenia (DITP); to an infectious cause due to the coexistence of neutrophilia; or related to thrombotic thrombocytopenic purpura (TTP), which could justify the liver involvement.¹⁵

The pathological mechanisms of SHH are complex, poorly understood and accepted to be multifactorial. Hepatic necrosis can coexist if there is severe liver involvement (best described in cases of HELLP syndrome) - being vasospasm, endothelial damage, microvascular thrombi with disseminated intravascular coagulation (DIC) and hypoperfusion hypothesized mechanisms that cause necrosis and rupture.¹⁰ Parenchymal compression by a large subcapsular hematoma can also be the cause of parenchymal necrosis. Subcapsular bleeding and hematoma usually precedes hepatic rupture, with insidious vague local or systemic symptoms followed by an acute phase of increasing pain and collapse.² In this specific case, an autoimmune undiagnosed disorder or a bleeding diathesis could be hypothesized as possible causes for bleeding. Bleeding diathesis from acute hepatic failure was excluded with normal values of PTT and INR;¹⁶ and thrombocytopenia was not sufficiently severe for this massive bleeding.

Due to its high mortality from intraabdominal massive haemorrhage and shock, and to the unspecific clinical manifestations (i.e. abdominal pain, nausea, vomiting and malaise and rarely shock) - imaging and laboratory evaluation play an important role in early diagnosis.^{1,4,5} Ultrasonography (US) and dynamic multidetector computerized tomography (MDCT) are the main diagnostic tools, and angiography is useful when there is also a therapeutic goal. Dynamic MDCT is the technique of choice due to its high accuracy, rapid acquisition and availability.^{4,5,9} It allows diagnosis, extension evaluation, location of foci of active bleeding and/or the determination of a possible underlying cause (i.e. tumour), detecting flow rates as low as 0.3mL/min.¹³ In some cases, an underlying cause may not be detected by cross-sectional imaging - due to diffuse parenchymal involvement or to massive haemorrhage obscuring focal parenchymal alterations.^{4,5,9}

Management is complex and should be individualized - mainly to clinical stability, the underlying cause and extent of bleeding, hepatic function and/or the existence of hepatic fibrosis and coagulation status. Haemostasis is the primary goal of treatment, firstly done by active resuscitation with fluids, blood product replacement and correction of coagulopathy.^{1,8,9} Conservative management may be appropriate if the patient is stable and the liver capsule is intact,² with few reports in the literature of stable patients with

SHH associated with HELLP syndrome - with small and/or contained haematoma - who were managed conservatively with imaging monitoring of stability.⁸ An emergent and multidisciplinary treatment is needed with unstable patients or when there are radiological signs of active bleeding. Interventional radiology management with transarterial embolization of selected vessels or one of the main hepatic branches or even the hepatic artery - are the preferred ways to primarily achieve haemostasis if possible.^{1,2,8,9} MDCT allows guidance of angiography and embolization. Angiography identifies foci of active bleeding of at least 0.5-1 mL/min. Haemostasis is restored by decreasing perfusion pressure and promoting clot formation after embolization.^{13,14} The used embolic agent depends largely on individual medical experience and preference; and coils are definitive agents that can be used safely in hepatic embolization due to the dual hepatic blood supply and consequently low risk of clinical significant parenchymal infarction. Immediate exploratory surgery - usually with perihepatic packing in the first step with or without plication and/or hepatic artery selective or nonselective ligation - may be needed due to hemodynamic instability, after failure of conservative and radiological intervention treatments with persistent or recurrent bleeding and/or in the setting of massive hepatic bleeding/necrosis; or when embolization is not feasible.^{1,4,5,8,9} A planned re-laparotomy can be done with appropriate and individualized further management.⁸ Hepatic resection - generally reserved for bleeding tumours in a second step and not to restore haemostasis after a carefully determination of staging and assessment of underlying liver function - was done successfully in selected cases on the acute phase in pregnant women and in patients with bleeding adenoma or HCC.^{1,2} Liver transplantation is an option when there is an uncontrolled haemorrhage or progressive fulminant liver failure.^{2,8} The management is even more complex if there is co-existing chronic liver disease, and those cases usually have poor outcomes.⁹ When haemostasis is achieved in the acute setting, subsequent treatment and outcome depend on the underlying condition.¹

Due to the difficulty to achieve diagnosis, this condition is associated with high mortality and strict therapeutic recommendations are lacking. Imaging and laboratory analysis play an important role in suspected cases.^{5,9} Outcome largely depends on the underlying pathology, extension and on the clinical background before the acute setting.¹

This rare condition with many underlying possible causes - in some cases obscure even after a careful clinical, imaging and pathological investigation - is a challenge in both diagnostic and therapeutic points of view, due to its rarity and complexity, incomplete understating and severity. Imaging plays a central role in the diagnosis and interventional radiology and surgery are cornerstones for management.

Acknowledgements

A special acknowledgement to Dr. Diogo Rocha, MD and Radiologist of Interventional Radiology of our Department, for his help in choosing the best images.

Received / Recebido 27/11/2017

Acceptance / Aceite 19/01/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial

References

1. Srinivasa S, Lee WG, Aldameh A, Koea JB. Spontaneous hepatic hemorrhage: a review of pathogenesis, etiology and treatment. International Hepato-Pancreato-Biliary Association Published Online First: 7 August 2015.
2. Mascarenhas R, Mathias J, Varadarajan R, Geoghegan J, Traynor O. Spontaneous hepatic rupture: a report of five cases. International Hepato-Pancreato-Biliary Association. Published Online First by Elsevier Inc.: December 2002.
3. Gelder HV, Gharibian N, Patel DB et al. Acute Haemorrhagic Myositis in Inflammatory Myopathy and Review of the Literature. Hindawi Publishing Corporation Case Reports in Rheumatology. Published Online First: 14 October 2014.
4. Furlan A, Fakhran S, Federle MP. Spontaneous Abdominal Hemorrhage: Causes CT Findings and Clinical Implications. AJR Am J Roentgenol. Published Online First: October 2009.
5. Klein K, Shapiro J. A. M. Spontaneous Hepatic Rupture with Intraperitoneal Hemorrhage without Underlying Etiology: A Report of Two Cases. International Scholarly Research Network ISRN Surgery. Volume 2011, Article ID 610747.
6. S Abraham, S Begum, D Isenberg. Hepatic manifestations of autoimmune rheumatic diseases. Ann Rheum Dis. 2004;63:123-9.
7. Soultati A, Dourakis S. Hepatic manifestations of autoimmune rheumatic diseases. Annals of Gastroenterology. 2005;18:309-24.
8. Wilson SG, White AD, Young AL, Davies MH, Pollard SG. The management of the surgical complications of HELLP syndrome. Ann R Coll Surg Engl. 2014;96:512-6.
9. Battula N, Tsapralis D, Takhar A, Coldham C. et al. Aetio-pathogenesis and the management of spontaneous hepatic bleeding in the West: a 16-year single-centre management. HEP. 2013;14:382-9.
10. Casillas V. J., Amendola M.A., Gascue A., Pinnar N. et al. Imaging of nontraumatic hemorrhagic hepatic lesions. RadioGraphics. 2000;20:367-78.
11. Cojocar M., Cojocar I. M., Silosi I., Vrabie C. D. Liver involvement in patients with systemic autoimmune diseases. MAEDICA – a Journal of Clinical Medicine. 2013;8:394-7.
12. Chen Zhe-Yu, Qi Qing-Hui, Dong Zuo-Liang. Etiology and management of hemorrhage in spontaneous liver rupture: a report of 70 cases. World J Gastroenterol. 2002;8:1063-6.
13. Ramaswamy RS, Choi HW, Mouser HC, Narsinh KH. et al. Role of interventional radiology in the management of acute gastrointestinal bleeding. World J Radiol. 2014;6:82-92.
14. Bauer JR., Jr. CER. Transcatheter arterial embolization in the trauma patient. Semin Intervent Radiol. 2004;21:11-22.
15. Izak M., Bussel J. B. Management of thrombocytopenia. F1000Prime Rep. 2014;6:45.
16. Bernal W, Wendon J. Acute liver failure. N Engl J Med. 2013;369:2525-34.
16. Rosales A, Que FG, Spontaneous hepatic hemorrhage: a single institution's 16-year experience. Am Surg. 2016;82:1117-20.