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Review Article / Artigo Revisão

Aggressive Fibromatosis of Desmoid Type - a Pictorial Review

Fibromatose Agressiva do tipo Desmoide - Revisão Pictórica

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Abstract

Deep fibromatosis of desmoid type comprises rare mesenchymal tumors characterized histologically by proliferation of fibroblasts and myofibroblasts. These lesions are characterized by infiltrative growth and local recurrence but an inability to metastasize.
They can be located in the abdominal wall (generally in sutures), intra or extraabdominal.
The best imaging modality for evaluation and staging of the deep fibromatoses is MR imaging. These well or ill-defined lesions generally present internal hypointense bands, with lack of enhancement in post contrast images (collagen bundles) and are usually centered in an intermuscular location with a rim of fat ("split fat sign"), although invasion of surrounding muscle is frequent. Linear extension along fascial planes ("fascial tail sign") is also a frequent manifestation in deep fibromatoses and is uncommon with other soft-tissue neoplasms.
MR image signal intensity has an implication on tumor recurrence, with a higher recurrence rate in lesions with high T2 signal. In surgically untreated lesions, with undergoing radiation or drug therapy, MR surveillance has been used to assess response to treatment with positive results demonstrating decrease in T2 signal, lesion enhancement and lesion size.

Keywords

Aggressive fibromatosis; desmoid tumor.

Resumo

As fibromatoses profundas do tipo desmoide constituem tumores mesenquimatosos raros que se caraterizam histologicamente por proliferação de fibroblastos e miofibroblastos. São caraterísti-cas destas lesões o crescimento infiltrativo e comportamento recidivante, sem capacidade de metastização.
Podem localizar-se na parede abdominal (geralmente em locais de suturas), ou em topografia intra ou extra-abdominal.
A melhor técnica de imagem para a avaliação e estadiamento das fibromatoses profundas é a RM. Estas lesões, bem ou mal definidas, exibem geralmente bandas internas de hipointensidade de sinal, sem captação de contraste (feixes de colagéneo) e encontram-se nos planos intermuscu-lares com um halo de gordura ("split fat sign"), embora invasão dos músculos adjacentes seja fre-quente.
Extensão linear ao longo dos planos fasciais (o "fascial tail sign") é igualmente uma manifestação frequente das fibromatoses profundas e incomum noutras neoplasias de tecidos moles.
A intensidade de sinal na RM tem implicação na recorrência tumoral, sendo que lesões com hipe-rintensidade de sinal em T2 têm maior probabilidade de recidiva. Na ausência de excisão cirúrgi-ca, em lesões em regime de radioterapia ou terapia farmacológica, a RM tem sido usada na ava-liação da resposta ao tratamento, com resultados favoráveis evidenciando diminuição da intensi-dade do sinal T2, da captação de contraste e da dimensão da lesão.

Palavras-chave

Fibromatoses agressivas; Tumor desmoide.

1. Introduction

1.1 Background

Deep fibromatoses (DF) of desmoid type, also known as desmoid tumors, are rare mesenchymal tumors characterized histologically by proliferation of fibroblasts and myofibroblasts.[1] They are also composed of dense deposits of intercellular collagen fibers and variable amounts of extracellular myxoid matrix and vessels.[2]
DF includes a large group of fibrous tissue proliferations that have intermediate biologic behavior between benign and malignant (fibrosarcoma) fibrous lesions. These lesions are characterized by infiltrative growth and local recurrence but an inability to metastasize.
Classically fibromatoses are divided into superficial (palmar, plantar and penile fibromatosis) and deep (DF of desmoid type) forms. (Fig. 1)
The main differential diagnosis of DF is malignant soft-tissue sarcoma, extranodal lymphoma, and benign myositis ossificans.

1.2 Deep Fibromatosis (DF) / Desmoid Type Fibromatosis

DF (desmoid type fibromatosis) typically manifests as a poorly circumscribed soft-tissue mass either in the abdominal wall, intra or extraabdominal in location.
This group of fibromatosis consists of rapidly growing lesions that often reach a large size and have a high tendency to recur after treatment, hence the term "aggressive fibromatosis".[3]

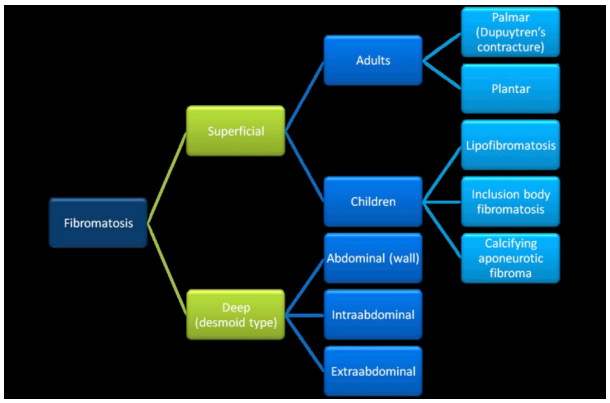

Figure 1 - Schematic illustration of the different types of fibromatosis: superficial and deep (desmoid) forms.

It tends to occur in patients in the 2nd to 4th decades of life, with a peak incidence between 25-35 years of age. Women are more commonly affected than men.
Slow insidious growth is common, and lesions are usually painless. Decreased mobility, reduced joint motion and neurologic complaints are less frequent symptoms. Nevertheless, most patients with DF are asymptomatic and these lesions are accidentally identified on imaging studies, most often on CT evaluations. (Figs 2, 3 and 4)
Most cases are sporadic, but there is a clear association with familial adenomatous polyposis and Gardner's syndrome, suggesting a link with mutations of the APC gene on chromosome 5q22. Approximately 10% of patients with Gardner syndrome also have fibromatosis, which typically is of the intraabdominal type, usually located in the small-bowel mesentery.[1]

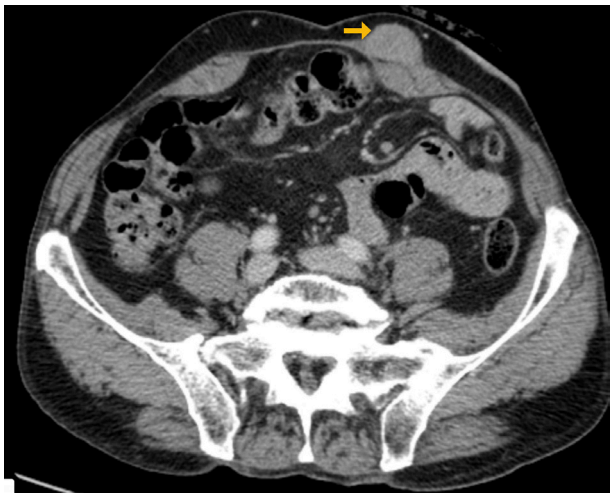

Figure 2 - Abdominal wall fibromatosis in a 64-year-old female. Axial CT enhanced image showing a well defined nodular enhancing lesion (arrow) in the left rectus abdominis muscle.

Intraabdominal DF may be either mesenteric, pelvic or retroperitoneal in locations, with similar appearances.
Extraabdominal DF can occur almost anywhere in the body, although they have a predilection for the upper torso including the upper arm (28%), chest wall/paraspinal (17%), and head/neck (10% to 23%). They are typically solitary lesions (85%) with a tendency to grow along fascial planes and to extend a great distance from the predominant mass.[4]
Abdominal wall fibromatosis is a distinct variant of the DF as it tends to occur in women during pregnancy or

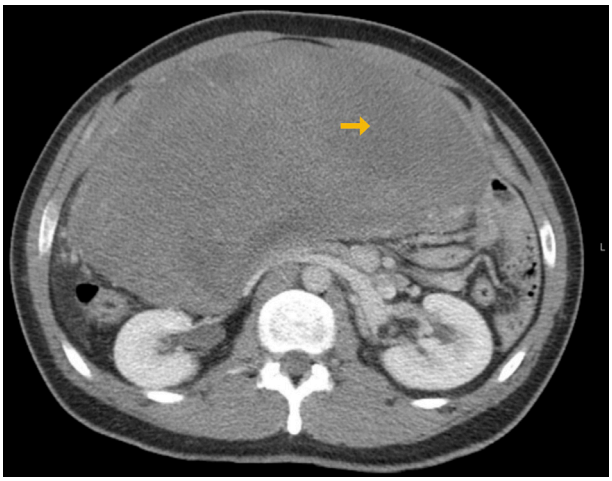

Figure 3 - Massive intraabdominal fibromatosis in a 45-year-old male. Axial CT image demonstrating a large heterogeneously enhancing intraabdominal mass. Note the areas of less contrast enhancement due to increase collagen content and myxoid degeneration (arrow).

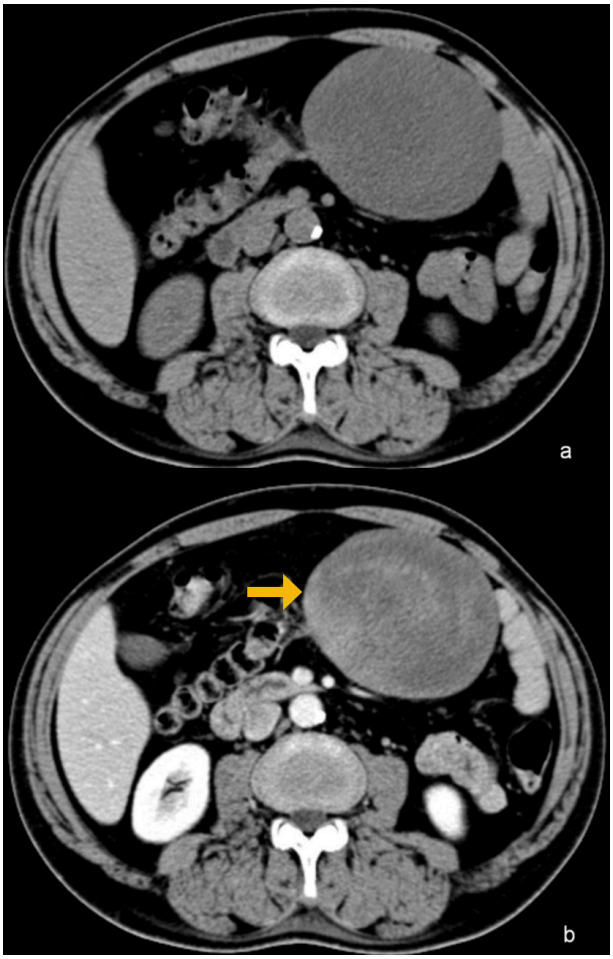

Figure 4 - Intraabdominal fibromatosis in a 39-year-old female. Axial non-enhanced (a) and enhanced (b) CT images showing a well defined, heterogeneously enhancing peritoneal mass. Note the well-defined borders of the lesion (arrow).

within the 1st year after delivery and in women who use oral contraceptives. The rectus abdominis and internal oblique muscles of the anterior abdominal wall are most frequently affected. (Fig. 5)

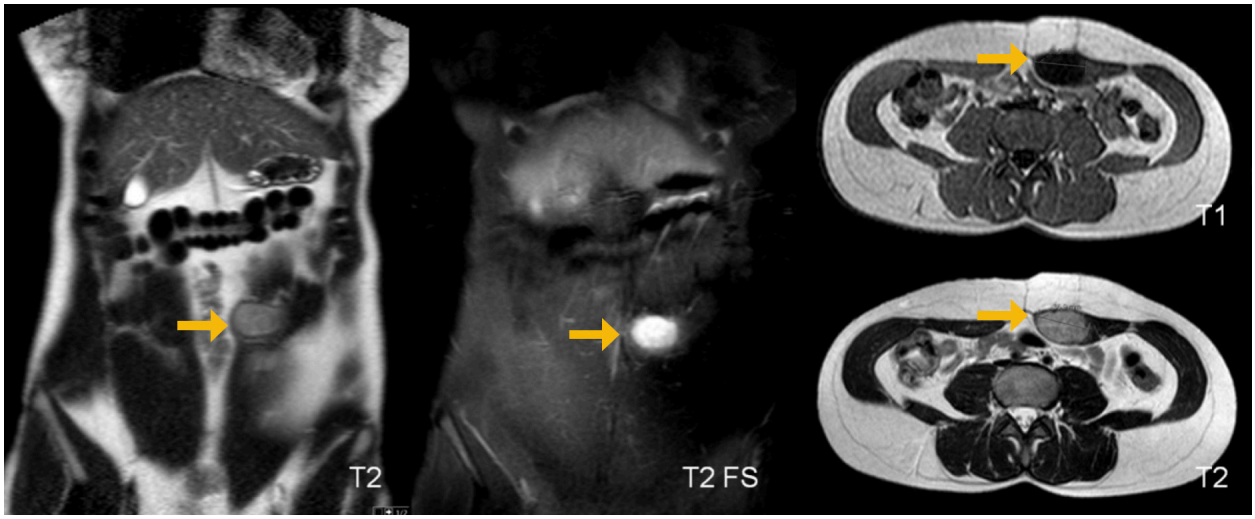

Figure 5 - Abdominal wall fibromatosis in a 37-year-old female. MR images showing a well defined nodular lesion in the left rectus abdominis muscle (arrows) with hypointense signal on T1-wheighted images, faint hyperintense signal on T2-weighted images and conspicuous hyperintense signal on fat saturated sequences.

2. Imaging Characteristics

2.1 Conventional Radiography

On conventional radiographs DF is either undetectable or identified as an amorphous soft-tissue-density mass. Calcification is uncommon. Bone involvement is noted in 6%-37% of cases more frequently after multiple recurrences.[7]

2.2 Ultrasound

Ultrasound evaluation reveals a hypoechoic lesion either ill or well-defined. Color Doppler evaluation is useful in demonstrating vascularity of these lesions although absent in 66% of cases.[7]

2.3 Computed Tomography

DF can be hypoattenuating, hyperattenuating or appear as whorled masses because of the alternating collagenous and myxoid areas. These lesions are well vascularized resulting in contrast enhancement on CT and MR studies, although the degree of contrast enhancement is variable.[7]

2.4 MR Imaging

MRI usually depicts a T1 and T2 intermediate signal intensity intermuscular centered lesion with a rim of fat ("split fat"sign), although invasion of surrounding muscle is frequent.

Linear extension along fascial planes (the "fascial tail" sign) is also a frequent manifestation (80%) and is uncommon in other soft-tissue neoplasms.[8] (Fig. 6 and 7)

In the majority (86%) of fibromatoses the T2-weighted images depict hypointense bands, with lack of enhancement in post contrast images, that are likely to correspond to dense conglomerations of collagen bundles, a feature that is more common in fibromatoses than in other neoplastic lesions, although not specific.[9,10,11] (Figs. 8, 9, 10, 11, 12, 13, 14 and 15).

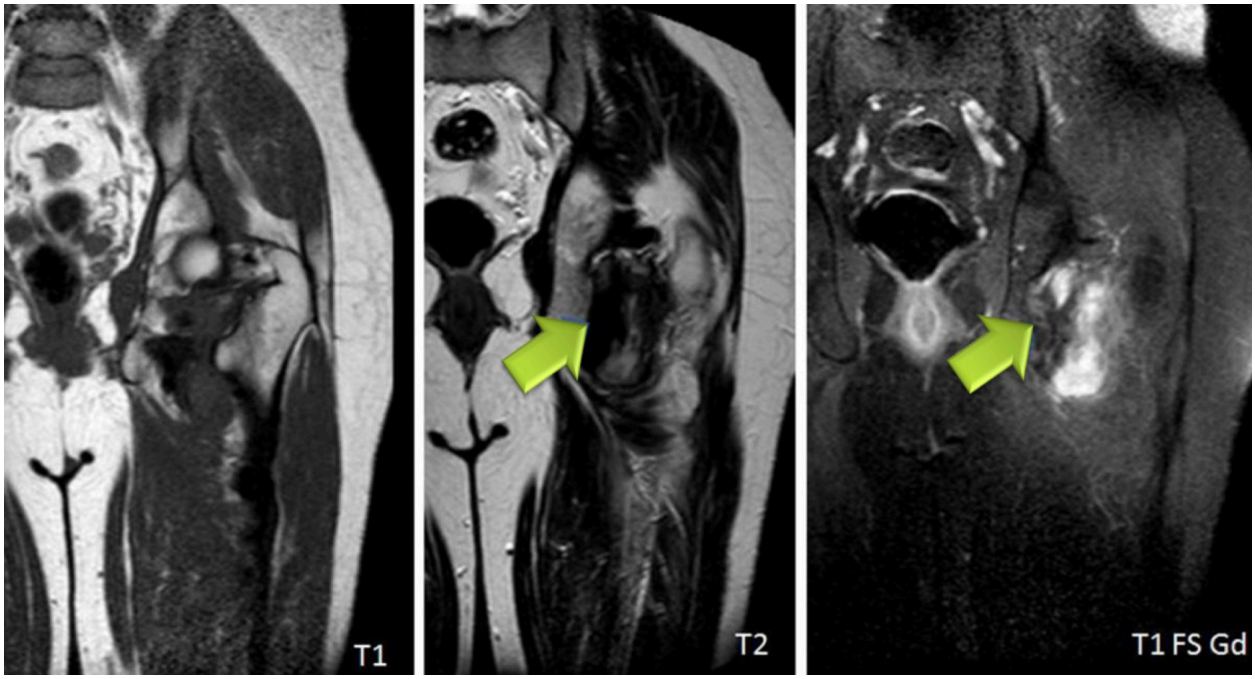

Figure 6 - Aggressive fibromatosis of the left upper thigh in a 44-year-old female. MR images showing an infiltrative heterogenous lesion with areas of low signal intensity in all pulse sequences (arrows). Following gadolinium administration these collagenized bands demonstrate lack of enhancement.

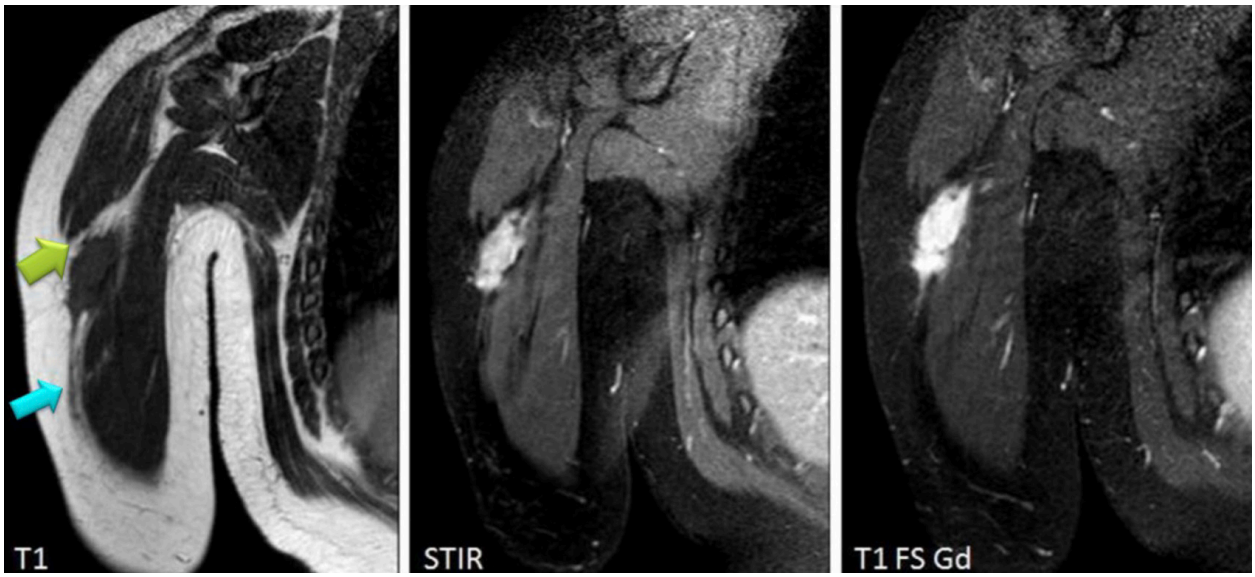

Figure 7 - Aggressive fibromatosis in the right upper arm in a 42-year-old female. The lesion is centered in an intermuscular location showing hypointense signal on T1-wheighted images, hyperintense signal on fluid sensitive sequences (STIR) and avid gadolinium enhancement. It shows rim of fat ("split-fat" sign, green arrow) and linear extension along fascial planes ("the fascial tail sign") (blue arrow).

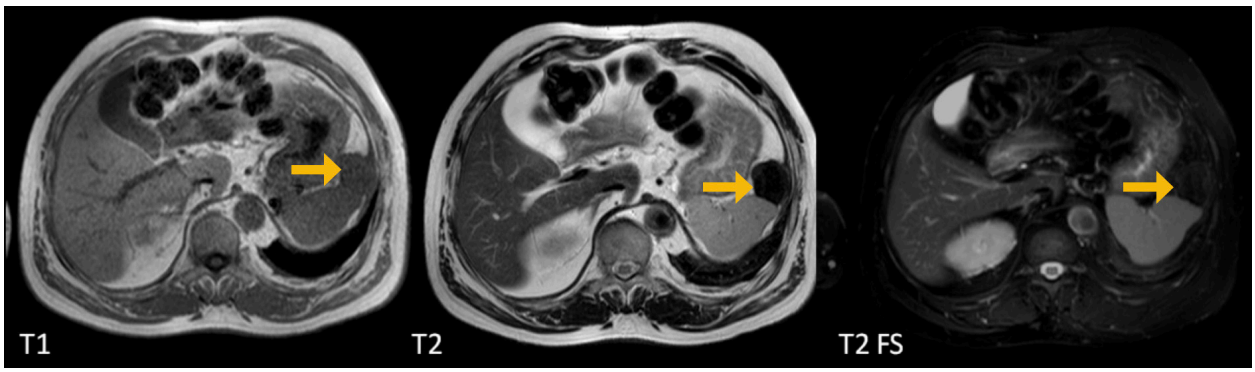

Figure 8 - Intraabdominal fibromatosis (mature form) in a 40-year-old female. MR images showing a well-defined intraabdominal mass anterior to the spleen (arrows) with intermediate signal on T1 weighted images and low signal on T2 and T2 FS weighted images.

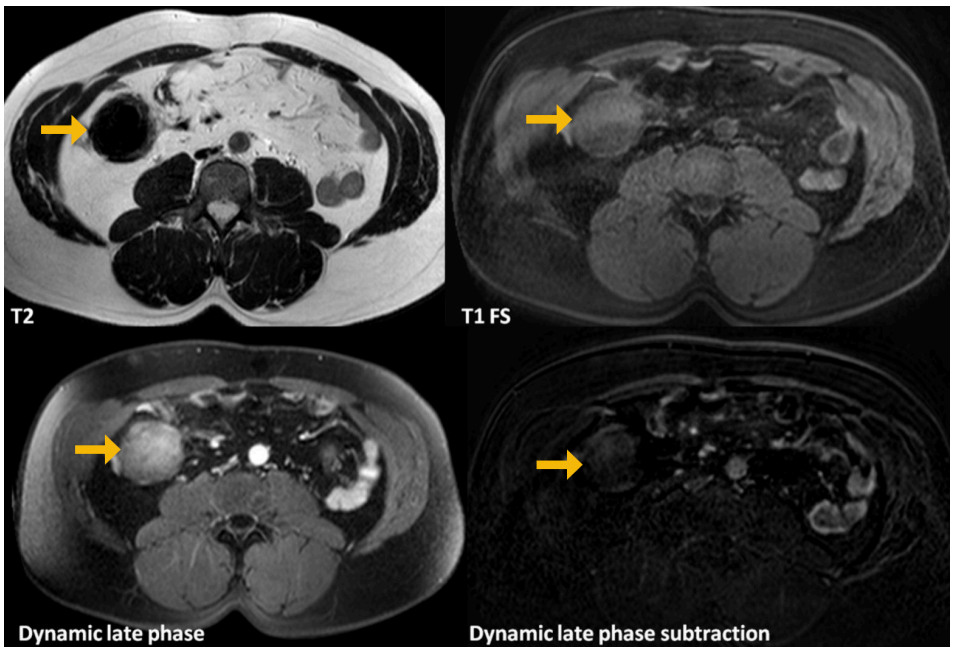

Figure 9 - Intraabdominal fibromatosis (intermediate form) in a 57-year-old male. MR images showing a well defined intraabdominal mass (arrows) with intermediate signal on T1 weighted images, low signal on T2 weighted images and slight contrast enhancement.

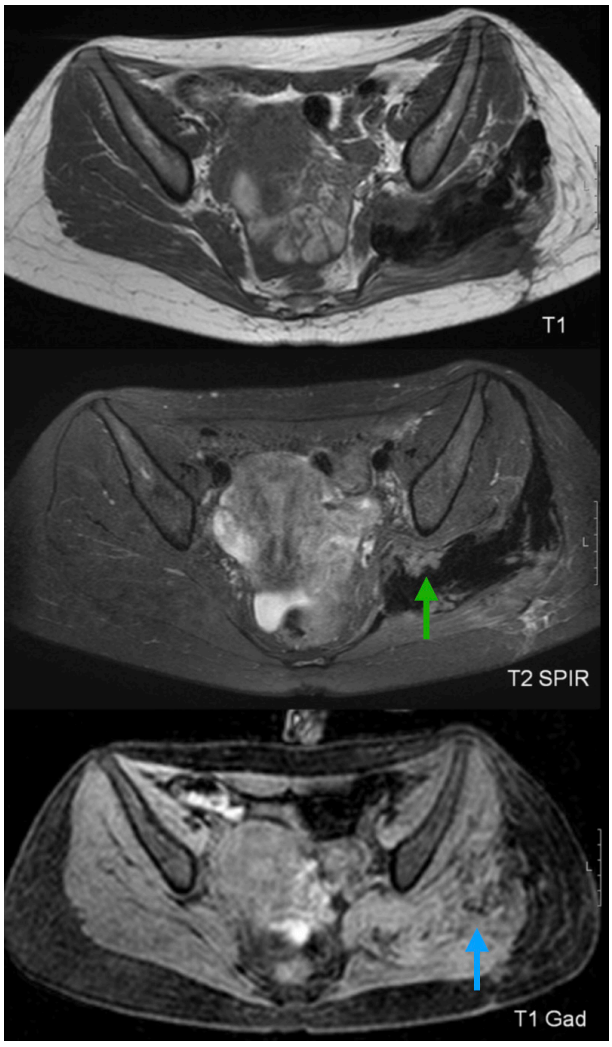

Figure 10 - Deep "active" fibromatosis of the left gluteus muscle in a 49-year-old male. Axial MR images showing an infiltrative mass in left gluteal region with heterogenous low signal intensity on T1-weighted and T2-weighted images. High intensity signal areas on T2-weighted images (green arrows) and avid enhancement (blue arrows) suggest "active"/first stage disease.

Desmoid type fibromatosis tumors have been described as having three stages of histologic evolution over time as they mature:[12]

- In the first stage, lesions are more cellular with large extracellular spaces (which appear as decreased T1 and increased T2 MR signal intensity). (Figs. 5, 10, 11, 12, 13 and 15)
- In the second stage, there is an increase in the amount of collagen deposition (which increases the heterogeneity of T2 signal) in the central and peripheral areas of the tumor. (Fig. 9)
- In the final stage there is an increase in the fibrous composition (which decreases the T1 and T2 MR signal intensity), with a decrease in the volume of the extracellular spaces and water content. (Figs 8 and 14)

3. Treatment

The preferred treatment is usually wide-local excision.
Adjuvant radiation therapy following surgery has been shown to decrease the local recurrence rate versus surgery alone.[13]
Additional therapies with reported positive results include radiofrequency ablation and chemotherapy agents. Several pharmacological treatments such as hormonal therapy (tamoxifen), nonsteroidal anti-inflammatory drugs (celecoxib), tyrosine kinase inhibitors such as imatinib or sorafenib and cytotoxic chemotherapy have been associated with clinical benefit in desmoid patients with progressive and/or recurrent disease. Antiestrogen (tamoxifen) and anthracycline-containing regimes appear to be associated with higher radiological response rates against desmoid tumors.[14]

4. Prognosis

Local recurrence rates are relatively high (50% of patients older than 20 years experiencing a recurrence after local excision).[15]
Signal intensity on long TR sequences may have an implication on tumor recurrence, with a higher recurrence rate in lesions with high T2 signal.

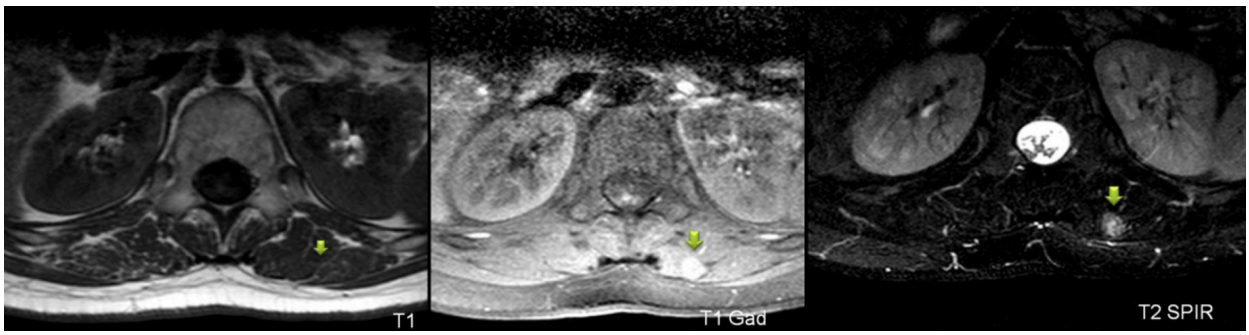

Figure 11 - Spinalis muscle fibromatosis in a 59-year-old female. MR images showing a well defined nodular lesion (arrows) in the left spinalis muscle with isointense to muscle signal on T1-wheighted images, faint hyperintense signal on T2-weighted images with fat saturation and avid contrast enhancement.

Although MR imaging can provide clues to the correct diagnosis of aggressive fibromatosis, histologic confirmation is required for a final diagnosis in all cases. To be noted that MR imaging features similar to fibromatosis may occur in pigmented villonodular synovitis, granular cell tumor, fibrosarcoma, and malignant fibrous histiocytomas.

In lesions undergoing radiation or drug therapy, MR surveillance has been used to assess response to treatment with a positive response demonstrating a decrease in T2 signal, lesion enhancement and lesion size.[16]

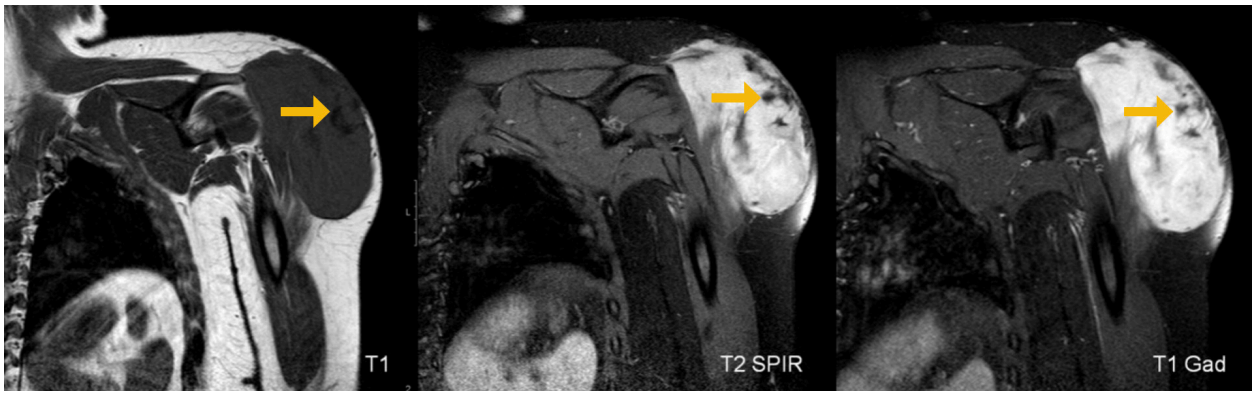

Figure 12 - Deep fibromatosis in the left deltoid muscle in a 54-year-old male. MR images showing a well defined mass with heterogenous intermediate signal intensity on T1-weighted images (isointense to the muscle), high signal intensity on T2- weighted images and avidly enhancing. Note the low signal intensity internal areas (arrows) with band like morphology demonstrating lack of enhancement (characteristic of fibromatosis).

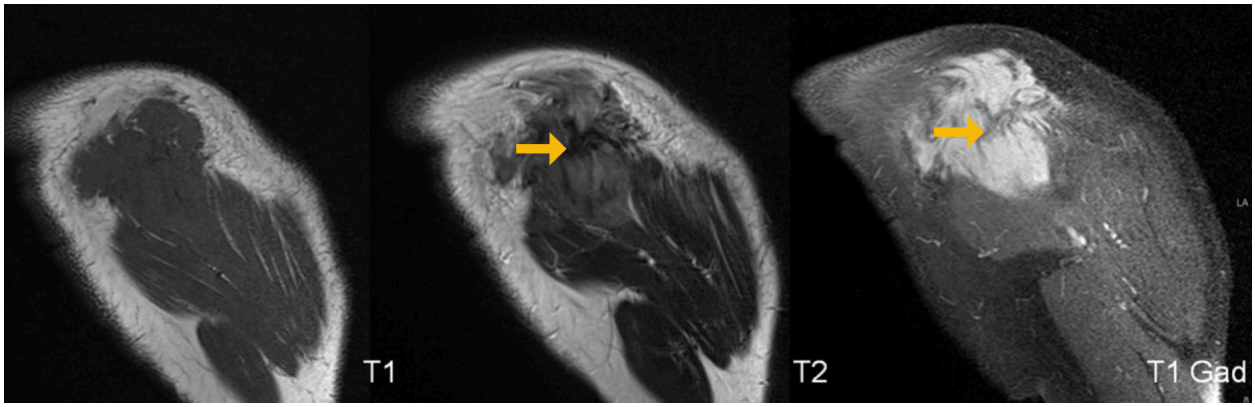

Figure 13 - Deep fibromatosis of the left deltoid in a 52-year-old female. Coronal MR images showing a mass centered in the left deltoid muscle with isointense signal on T1-wheighted images, faint hyperintense signal on T2-weighted images and avid contrast enhancement. Note the central T2 -weighted hypointense and non-enchancing collagen bands (arrows).

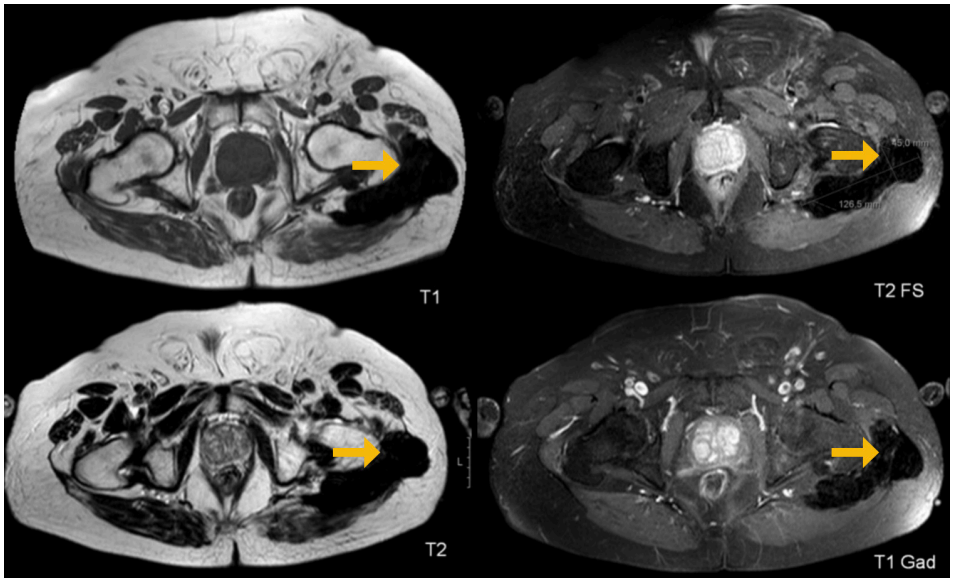

Figure 14 - Deep fibromatosis (late stage) of the left gluteal region in a 34-year-old female. MR images showing an infiltrative mass centered in the left gluteus muscles (arrows) with low signal intensity in all pulse sequences. Following gadolinium administration it demonstrates lack of enhancement.

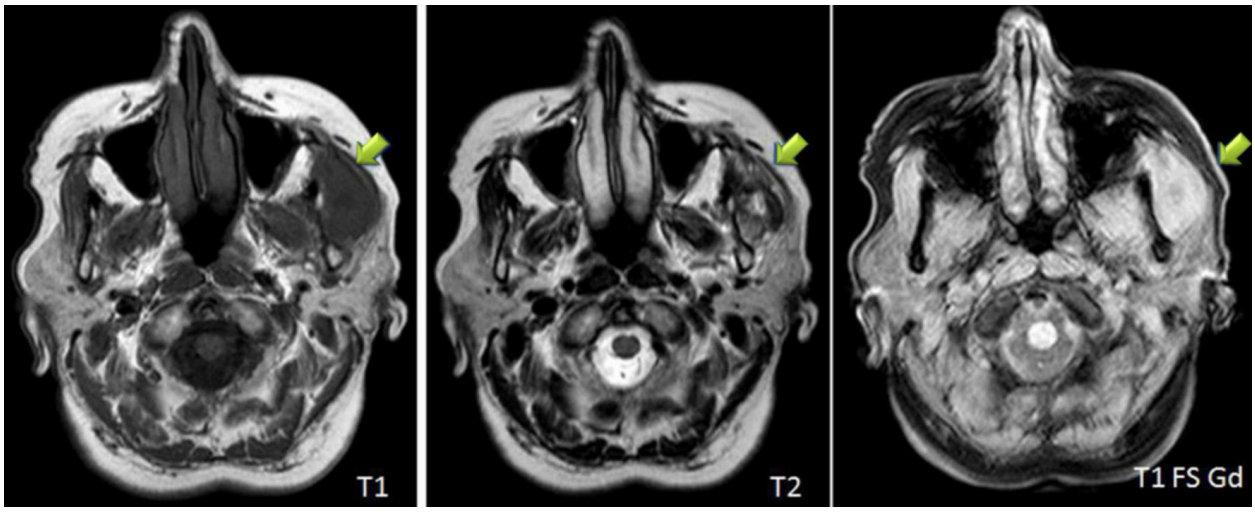

Figure 15 - Deep fibromatosis of the left masseter muscle in a 43-year-old male. Axial MR images showing a isointense to muscle lesion on T1 weighted images, hiperintense in T2weighted images and avid contrast enhancement lesion on the left masseter muscle (arrows). It was biopsy proven to be a deep fibromatosis.

5. Conclusion

AF should be included in the differential diagnosis of a soft-tissue mass.
MRI has come to play a crucial role in the management of AF, from diagnosis to follow-up. The deep fibromatoses are often heterogeneous with internal T2-weighted hypointense bands. The "fascial tail" sign and "split fat" sign add diagnosis specificity to MR imaging.
MR imaging can also be useful for determining the response to adjuvant radiation therapy or chemotherapy - responding lesions show lower T2 signal intensity, decreasing contrast enhancement and decreasing dimensions.

6. Main Points

- Fibromatosis can be superficial (palmar, plantar) or deep/ aggressive fibromatosis of desmoid type (either in abdominal wall, intraabdominal or extraabdominal locations).
- Desmoid type fibromatosis are mesenchymal tumors with high tendency to recurrence.
- While extraabdominal forms can occur almost anywhere in the body, they have a predilection for the upper torso, including the upper arm, chest wall/paraspinal, and head and neck locations.
- The best imaging modality for evaluation and staging of the deep fibromatoses is MR imaging.
- Deep fibromatoses may be depicted in MR image with non-enhancing T2-weighted hypointense internal bands that correspond to collagenous regions seen at histologic analysis, with the presence of "split fat"sign and fascial tail sign.
- The signal intensity on T2 MR image is a prognostic factor as there is higher recurrence rate in lesions with high T2 signal.
- MR imaging can be useful for determining the response to adjuvant radiation therapy or chemotherapy - responding lesions show lower T2 signal intensity, decreasing contrast enhancement and decreasing dimensions.

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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).
Protecção 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.

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