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Radiological Case Report / Caso Clínico

Infantile Hemangioma: A Case Report

Hemangioma Infantil: A Propósito de um Caso Clínico

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Abstract

The authors present a case report of a newborn, admitted for pre-auricular swelling associated with a bright red skin patch. The ultrasound and magnetic resonance imaging evaluations were compatible with the diagnosis of parotid gland infantile hemangioma. Infantile hemangioma is a common benign vascular tumor, representing the vascular anomaly most frequently found in pediatric age. Its differential diagnosis includes other vascular tumors and vascular malformations. In most cases, expectant observation is recommended, with pharmacological and / or surgical treatment reserved for particular cases.

Keywords

Hemangioma / diagnostic imaging; Vascular malformations; Vascular neoplasms; Newborn.

Resumo

Os autores apresentam o caso clínico de um recém-nascido, admitido por tumefação pré-auricular associada a mancha cutânea com coloração vermelho vivo. As avaliações imagiológicas por ecografia de partes moles e ressonância magnética (RM) foram compatíveis com o diagnóstico de hemangioma infantil parotídeo. O hemangioma infantil é um tumor vascular benigno comum, representando a anomalia vascular mais frequentemente encontrada em idade pediátrica. O seu diagnóstico diferencial engloba outros tumores vasculares e malformações vasculares. Na maioria dos casos é recomendada observação expectante, sendo o tratamento farmacológico e/ou cirúrgico reservado para casos particulares.

Palavras-chave

Hemangioma/diagnóstico por imagem; Malformações vasculares; Neoplasias vasculares; Recém-nascido.

Introduction

Infantile hemangioma represents the vascular "anomaly" most frequently found in pediatric age.[1-3] It is not usually present at birth, with onset in the first weeks/months of life. The International Society for the Study of Vascular Anomalies (ISSVA) classifies infantile hemangioma as a vascular tumor,[4] due to the presence of endothelial cell proliferation. The diagnosis is generally clinical and the image allows assessing the extent of the tumor and its relationship with neighboring structures, a particularly important aspect in locations with a higher risk of complications.
The authors describe a clinical case of infantile parotid hemangioma in a newborn. The diagnosis evaluation and the relevance of complementary exams are discussed, as well as the therapy and follow-up strategy.

Clinical Case

Caucasian newborn, hospitalized for right preauricular swelling, appearing on the 22nd day of life and progressive volumetric increase.
During observation, a change in the bright red, protuberant and compressible skin color was documented (Fig. 1), with no other changes in the objective examination. The summary laboratory evaluation showed no changes.
For clarification, an ultrasound evaluation by ultrasound and MRI of the neck were performed.

The soft tissue ultrasound demonstrated the presence of a heterogeneous, well-defined mass, with lobulated contours, corresponding to the parotid gland with alteration of its echostructure and volume (Fig. 1), without extension or invasion of the adjacent planes subject to evaluation, intensely vascularized (Fig. 2), with large-calibre arterial and venous vessels inside.
At MRI, an asymmetry in the volume of the parotids was identified, seeing a marked increase in the right one, due to a lesion in its superficial and deep lobes, with regular contours and well-defined limits, with a larger axis of 48 mm (Fig. 3 and 4). The extension of the lesion to the deep lobe of the right parotid gland determined medial deviation of the parapharyngeal fat, without repercussions

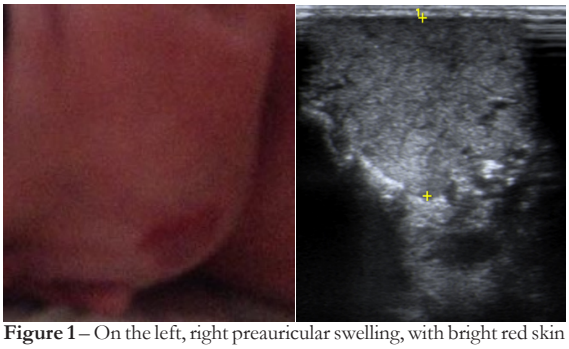

Figure 1 – On the left, right preauricular swelling, with bright red skin changes. On the right, a corresponding ultrasound image showing a heterogeneous, well-defined mass, with lobulated contours, in the topography of the parotid gland.

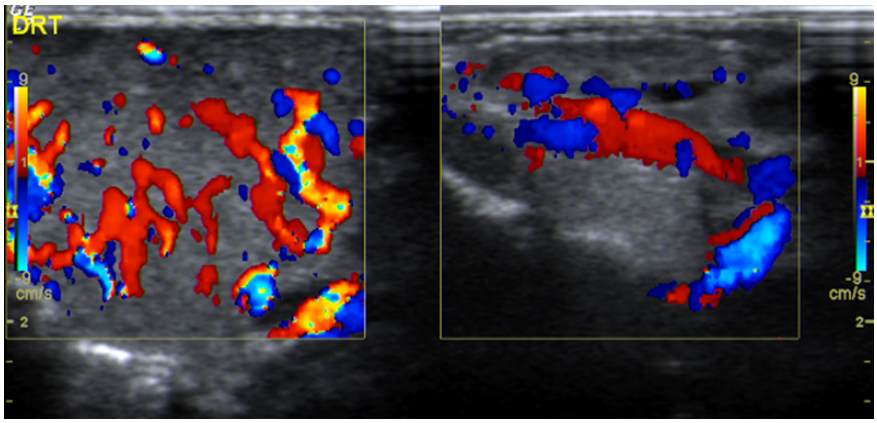

Figure 2 – Soft tissue ultrasound with Doppler: intensely vascularized mass, with large calibre vessels inside.

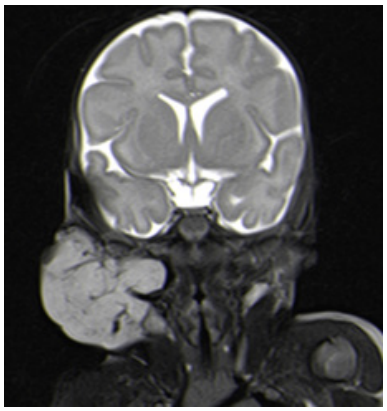

Figure 3 – MRI, coronal section with weighting in T2 FS: marked increase in the right parotid, due to a lesion affecting its superficial and deep lobes, well defined and with regular contours.

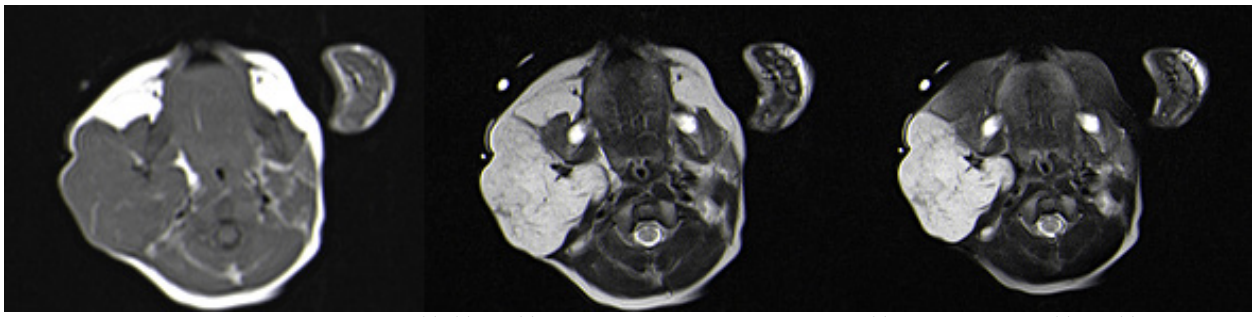

Figure 4 – MRI, axial sections, with weightings in T1, T2 and T2 FS: translated by intermediate signal in T1 and hypersignal in T2 and T2 FS, being possible to individualize some signs of vascular void inside. Lesion extension to the deep lobe of the right parotid gland determined medial deviation from homolateral parapharyngeal fat.

on the contour of the pharyngeal air column. Although the semiological characteristics by image of the right parotid lesion were not specific, given the clinical characteristics of the cutaneous alteration present in the same region, the most likely diagnostic hypothesis of mixed infantile hemangioma was considered. In the summary assessment of the encephalic parenchyma obtained, no malformative aspects were identified, particularly the posterior fossa, nor suspicious aspects of intracranial hemangiomas.
The study was complemented by abdominal ultrasound, echocardiogram and ophthalmic evaluation.
Abdominal ultrasound showed no changes.
The echocardiogram showed a 5 mm nodule adhering to the mitral valve cord and a 3 mm nodule in the left atrium. The short-term evaluation after therapy with propanolol showed resolution of these changes, suggesting that they were hemangiomas.
The ophthalmological evaluation did not reveal any changes.
Therapy with propanolol 3 mg/kg/day and prednisolone 4 mg/kg/day was performed, with a decrease in the volume and consistency of the lesion, in re-evaluation at 4 months of age, confirming the initial diagnosis. In the 6-month reassessment, the aim was to regress the cutaneous component, however, with a volumetric increase of the deep component, having proceeded with a therapeutic adjustment.

Discussion

Cervicofacial swelling is a common clinical problem in the pediatric population. It may have congenital or acquired causes, such as infection and benign or malignant tumors of the soft tissues and/or of the bone.
Clinical history and objective examination are the most important factors in diagnosis. In the presence of a slow-growing swelling, as in the case presented, the diagnostic hypotheses to consider are the presence of a mass, such as a neurofibroma, a vascular or lymphatic malformation, a hemangioma or a bone disease, such as fibrous dysplasia.
The presence of a protruding, compressible skin lesion with a bright red color, described as a "strawberry lesion",

raises the diagnostic hypothesis of infantile hemangioma as the most likely.

Infantile hemangioma is the most common vascular "anomaly" found in children[1,2,3] with a predisposition for females (3:1 ratio). It presents a characteristic life cycle: it is not usually present at birth, with onset in the first weeks/months of life and subsequent gradual volumetric increase (proliferative phase) and involution in the following years (involution phase), remaining as a variable amount of fibro-adipose tissue.[1-3] Intraparotid location is not frequent, but it is the most common benign tumor of the salivary glands in children.[5]

In certain locations, the hemangioma may have the potential to interfere with a vital structure or function, as is the case of airway, peri-orbital, liver, gastrointestinal tract or bulky and rapidly growing skin lesions.[6] Furthermore, in some locations, there is an increased risk of ulceration, residual skin changes and disfigurement, as it is the case with bulky, segmental or nodular hemangiomas and those located on the lips, nose or pinna.[7]

Infantile hemangioma may be part of the PHACE syndrome,[8] characterized by changes in the posterior fossa, head and neck hemangiomas, arterial changes, heart defects or coarctation of the aorta, ocular or endocrine changes and malformations of the sternum.

In the presence of a single lesion with typical clinical history and objective examination, complementary diagnostic studies, either imaging or histological ones, are not necessary. However, in specific cases, namely due to its topography and/or extension and suspected PHACE syndrome, imaging studies may be indicated for the correct characterization of the extension and its relationship with adjacent structures. The image also allows to classify the lesion in cases where clinical doubt persists.

Ultrasound and MRI are the two imaging modalities that allow obtaining information about blood flow and morphological characteristics, in order to obtain a differential diagnosis for this type of alteration.[9,10]

Ultrasound is the first-line imaging method, although it has some limitations. It has a limited spatial reach, preventing evaluation of lesions deeply located. In the ultrasound study, infantile hemangioma usually has an oval morphology and heterogeneous echostructure. The spectral Doppler reveals vessels with low resistance arterial curve.[3,11,12] With the involution process, an increase in the echogenicity of the lesion is observed, with decreased vascularization and increased vascular resistance.[3]

Extensive or complicated hemangiomas require MRI assessment to characterize and determine their extension.[13,14] Computed tomography has a limited role. Its main advantage lies in the speed of acquisition, useful in urgent situations.[15]

In MRI, in the proliferative phase, infantile hemangioma is a lobulated, well-defined lesion, with T2 hypersignal and several internal "signal void" artifacts. It has early and homogeneous enhancement after contrast. In the involution phase, there is a gradual decrease in enhancement and an increase in the fibroadipose component.[16]

Regarding differential diagnosis of this type of lesions, the ISSVA classification divides vascular anomalies into two groups: vascular or vasoproliferative tumors and vascular malformations.[4] Vascular tumors are characterized by the presence of endothelial cell proliferation, where vascular malformations are vascular structural changes in the arterial, venous, lymphatic or capillary system, not associated with proliferation of endothelial cells.

Infantile hemangioma, as well as congenital hemangioma, are both vascular tumors. They are distinguished by the presence or absence of the endothelial cell glicose transporter 1 (GLUT1) protein.[17,18,19]

Congenital hemangioma has a different life cycle from the infantile hemangioma. It develops in the fetal period and is fully developed at birth. It may subsequently undergo rapid involution after birth (rapidly involutive hemangioma) or remain unchanged over the years (non-involutive hemangioma).[20] The imaging findings of congenital hemangioma are similar to those seen in infantile hemangioma.[21]

Vascular malformations occur with equal frequency in both sexes and in 90% of the cases they are present at birth.[22] They show proportional growth to the child's growth. They are not associated with spontaneous regression, nor do they respond to corticosteroid treatment. They are divided into low-flow lesions, including variable combinations of lymphatic, capillary and venous elements, and high-flow lesions, which contain an arterial component (arteriovenous shunts). They are characterized by an agglomeration of vascular channels of altered morphology, without interposed stroma.[22]

High-flow vascular malformations present increased arterial flow, similar to infantile hemangioma. However, some characteristics allow it to be distinguished. On ultrasound, high-flow vascular malformations appear as a cluster of vessels, with no interposed solid tissue. In the Doppler evaluation, there is arterial and venous flow, but there is arterialization of the venous flow curves. In MRI, they are characterized by serpiginous "signal void" artifacts, without solid tissue interposed, both in T1 and T2, corresponding to dilated and dysplastic arteries that drain into arterialized veins.[23,24]

Low-flow vascular malformations contain venous, lymphatic vessels or both.[21] They commonly manifest before the age of 2 and their most frequent location is the head and neck.[13] Venous malformations, sometimes referred to as "port wine spots", due to their color on objective examination, also represent an important differential diagnosis. On ultrasound, they are solid, hyperechogenic lesions, containing phleboliths and venous flow or absence of flow in the Doppler evaluation. On MRI, they have an intermediate and heterogeneous signal at T1, hypersignal at T2 and solid areas with contrast uptake.[21] Lymphatic malformations are distinguished by being multiquistic, with or without liquid-liquid levels. They do not present vascular flow, except in the septa.[21] Venolymphatic malformations have combined characteristics.

Once the diagnosis of infantile hemangioma has been established, expectant observation is recommended in most cases, since most regress spontaneously and are not associated with complications. In some cases local complications, such as ulceration, and systemic complications occur, such as high-output heart failure and respiratory compromise.[25,26] In the presence of complications or risk of disfigurement, there is an indication for active therapy, which includes the administration of anti-angiogenic drugs such as propanolol and corticosteroids and, in some cases, laser therapy, surgery and embolization.[25]

Final Considerations

Infantile hemangioma represents the most common vascular tumor in pediatric age. Imaging complements clinical evaluation, allowing to clarify the diagnosis and evaluate the extent of the tumor and its relationship with neighboring structures. Other vascular tumors and vascular malformations are part of the differential diagnosis. The correct diagnosis allows targeted treatment.

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Ethical disclosures / Divulgações Éticas

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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).
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