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Radiological Case Report / Caso Clínico

Angiomatoid Fibrous Histiocytoma: A Rare Tumor and a Diagnostic Challenge

Histiocitoma Fibroso Angiomatóide: Um Tumor Raro e um Desafio Diagnóstico

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Abstract

Angiomatoid Fibrous Histiocytoma (AFH) is a rare soft tissue neoplasm that often presents in the subcutaneous or deep dermis of the extremities of children and young adults and is classified according to the WHO Classification of Soft Tissue Tumors 2020 as "intermediate tumor of uncertain differentiation". Diagnosing this condition is challenging for radiologists and pathologists because of its nonspecific and heterogeneous imaging and histopathologic features.
The authors present a case of a 13-year-old girl with a painless, tender, and nonmobile, slow-growing palpable lump in the right thigh, that on MRI was well-defined, with pseudocapsule and lobulated contours, with fluid-fluid levels with hemorrhagic content and a solid enhancing nodule. Due to the nonspecific imaging features favorable to a malignant lesion, a percutaneous biopsy was performed, and the result was inconclusive. The final diagnosis of AFH was made after surgical excision of the lesion.

Keywords

Soft tissue neoplasms; Angiomatoid fibrous histiocytoma; Pediatrics; Musculoskeletal.

Resumo

O Histiocitoma Fibroso Angiomatóide (AFH) é uma neoplasia de tecidos moles rara que se apresenta na espessura do tecido celular subcutâneo e derme profunda das extremidades, em crianças e jovens adultos, e é classificado segundo a Classificações de Tumores de Tecidos Moles de 2020 da OMS como "tumor de grau intermédio de diferenciação incerta". Pelas suas características inespecíficas, o diagnóstico desta condição é desafiante tanto para os Radiologistas como para os Patologistas.
Os autores apresentam o caso de uma rapariga, 13 anos, com uma lesão expansiva de crescimento lento na coxa direita, não dolorosa, dura e não mobilizável, que no estudo de RM correspondia a uma lesão bem definida, com pseudocápsula e contornos lobulados, com nível de líquido hemorrágico e um nódulo sólido com realce após contraste i.v. Pela suspeita de lesão maligna foi realizada uma biópsia percutânea cujo resultado foi inconclusivo. Após recessão cirúrgica da lesão foi feito o diagnóstico definitivo de AFH.

Palavras-chave

Neoplasia de tecidos moles; Histiocitoma fibroso angiomatóide; Pediatria; Musculoesquelético.

Case Report

A 13-year-old girl presented with a painless, tender, and nonmobile, slow-growing palpable lump in the anteromedial middle third of the right thigh, without associated cutaneous findings. Personal and familiar clinical history was unremarkable, and systemic signs or symptoms were negative.
Imaging evaluation was made with MRI, which showed a heterogeneous mass in the middle third of the thigh, along the medial and vastus intermediate muscles and the distal part of the adductor longus, measuring approximately 5cm. It was a well-defined mass, ovoid in shape, with pseudocapsule and lobulated contours. The lesion presented a solid nodular portion measuring around 3cm, with an intermediate T2 signal similar to the adjacent muscle on T1, which was avidly enhanced after gadolinium injection. This solid part was surrounded by a liquid area, with fluid-fluid levels and a hematocrit effect in relation to hemorrhagic content. The lesion was in strict contact with the vascular and nervous saphenous structures, causing compression and, possibly, adherence. There was no extra-fascial or osseous invasion. (Figure 1)

Although nonspecific, the imaging features favored an aggressive lesion, especially for the presence of a soft tissue enhancing nodule, and in this age group, the hypothesis of synovial sarcoma was considered the most likely.
A computed tomography (CT) of the neck, thorax, abdomen and pelvis was performed without signs of secondary disease, and an 18F PET/CT revealed a heterogeneous hypermetabolism also suspected for tumoral malignant lesion.
For this reason, an ultrasound-guided percutaneous biopsy was performed, but it was not conclusive due to insufficient samples.
After wide surgical resection, the diagnosis of Angiomatoid Fibrous Histiocytoma (AFH) was made.

Discussion

Angiomatoid Fibrous Histiocytoma (AFH) is a rare, slow-growing, soft-tissue mesenchymal neoplasm, previously known as Angiomatoid Malignant Fibrous Histiocytoma, as one of the five subtypes of Malignant Fibrous Histiocytoma variants.[1] Nonetheless, AFH is no longer classified as malignant because of its benign microscopic appearance and

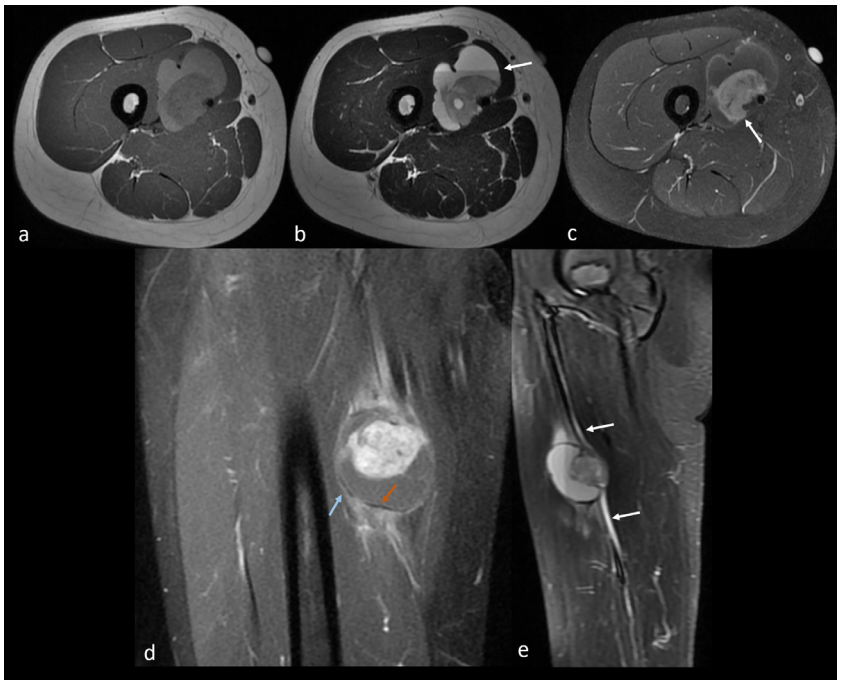

Figure 1 – MRI axial T1-weighted (a), axial T2-weighted (b), axial (c) and coronal (d) T1-weighted with fat saturation after administration of endovenous gadolinium-based contrast agent and sagittal STIR (e). Nodular lobulated lesion in the anteromedial middle third of the right thigh, with pseudocapsule and mixed solid and cystic component, with fluid-fluid level with hematocrit effect (hemorrhagic content) (arrow in b), and a nodular solid part that enhanced after endovenous contrast administration (arrow in c). The "double rim sign" was present in the periphery of the lesion, with a superficial enhancing rim (blue arrow in d) and an inner hypointense rim (orange arrow in d). On sagittal STIR imaging, the close relation with the saphenous neurovascular bundle was documented (arrows in e).

favorable prognosis and nowadays is classified according to the WHO Classification of Soft Tissue Tumors 2020 as an "intermediate tumor of uncertain differentiation".[2]

AFH usually presents in the subcutaneous or deep dermis of the extremities, even though it may occur anywhere in the body, with cases described in the head and neck and trunk region. It is more common in children and young adults, with only a few rare instances identified in older people.[1,3] Although it is generally a superficial lesion, in around 18% of the cases, it may invade deep structures such as skeletal muscle or periosteum.[4]

The clinical behavior of this entity is very nonspecific, usually presenting as a slow-growing painless mass, in rare cases with systemic signs and symptoms similar to paraneoplastic syndromes, such as fever, anemia or malaise, which make the clinical suspicion of this diagnosis very unlikely.[2] This indolent clinical presentation makes it misdiagnosed as hematoma or hemangioma.[5]

Due to its rarity, there is limited published literature about AFH, especially in the radiology literature, so its imaging features are not well established. Most AFH documented in the literature presents a combination of cystic and solid content, with the cystic part filled with blood products in relation to characteristic angiomatoid spaces of this tumor.[1] This is one of the reasons AFH is often challenging to differentiate from vascular tumors, such as hemangioendotheliomas, angiosarcomas or even organized hematomas.

Additionally, AFH is usually homogeneously isointense to muscle on T1-weighted sequences, although some may be hyperintense due to methemoglobin content. On T2-weighted imaging, AFH is typically heterogeneously hyperintense, and some mild hyperintensity in peritumoral tissues due to edema may be found.[1,6]

Martinez et al. described two standard imaging features of AFH. The first is the "double rim sign", present both on T2-weighted and post-contrast imaging, which consists of the presence of a superficial rim of high signal intensity, probably due to an inflammatory layer of lymphocytes and plasma cells, adjacent to a deeper rim of low signal intensity, rich on fibrous tissue, hemosiderin or undergoing hyalinization.[1,7]

Although most of the tumors have well-defined margins, Martinez et al. also described another typical infiltrative pattern, breaching the margin of the lesion and invading surrounding fat or muscle in a confluent or cord-like appearance.[1]

Additional findings, such as fluid-fluid levels, pseudocapsule, and multilocularity, are frequently present in imaging findings. Almost all AFH described in the literature enhanced after gadolinium administration, whether internal or nodular peripheral. The presence of hypointense nodular foci on T1 and T2-weighted sequences due to hemosiderin may be documented.[6]

This wide variety of imaging features described above may be present in an innumerable soft tissue lesion, such as traumatic, hemorrhagic or vascular conditions, as well as benign and malignant neoplasms. In the presence of those features associated with the "double rim sign" or tumor invasive margins, AFH should be added as a differential diagnosis, especially when present in the extremities of a pediatric or young adult patient.[1]

It is not rare that percutaneous needle biopsy doesn't provide a definite diagnosis of this entity, often misdiagnosed as other soft tissue tumors, such as myxoid liposarcoma, Ewing's sarcoma, myxofibrosarcoma or synovial sarcoma. For this reason, wide surgical excision and histopathologic analysis may be needed for the correct diagnosis, although its variable histomorphology is also a diagnostic challenge for pathologists. Classic spindle cell morphology of the tumor cells was the most common microscopic feature (76%), followed by epithelioid morphology (20%). However, there was an overlap of these two morphologies in 12% of the cases.[8]

Immunohistochemistry and genetic analysis will improve the accuracy of this entity's diagnosis. The presence of SOX9, a transcription factor involved in the embryogenesis of cartilage and gonadal tissue, is specific of AFH and has been expressed in 85% of the cases.[9]

Wide surgical excision is the standard treatment option for AFH. After excision, AFH usually develops an excellent clinical outcome and prognosis, with less than

1% of metastatic cases and a meager mortality rate.[5] Local recurrence occurs in 15% of patients after excision. For this reason, clinical and radiological follow-up is mandatory.[6]
Chemotherapy and radiotherapy are rarely used unless in the event of distant metastasis and multiple local recurrences.[7]
The absence of a pseudocapsule is favorable for tumor, inflammatory, blood cells and hemosiderin to extend beyond the apparent margins of the tumor to the adjacent tissues, and this is believed to be a positive factor for tumor recurrence. Additionally, some authors defend that a head and neck location and the depth of tumor extension are also correlated with aggressive behavior due to the increased difficulty in performing wide surgical excision, with an increase in local and distant metastasis on follow-up examinations.[5,10]
Imaging diagnosis of local recurrence is challenging because it may resemble post-operative changes, and the presence of local cystic nodules after tumor excision on follow-up examinations should raise suspicion of tumor recurrence.[1]

Conclusion

AFH is a rare soft tissue tumor, classified as an "intermediate tumor of uncertain differentiation" by the WHO Classification of Soft Tissue Tumors 2020. It is difficult to diagnose because of the lack of specific clinical, radiological, and histological features, showing a wide variety of morphologic imaging presentations, which frequently confuses it with malignant and benign conditions and mimics several other tumors.
The awareness of the radiologists for this diagnosis and the recognition of its more frequent imaging findings improve the chance of suggesting this diagnosis preoperatively, allowing complementary histopathological examination and wide surgical resection.

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.
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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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