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A rare case of cystic variety of angiomatoid fibrous histiocytoma

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Abstract. – A rare case of cystic angiomatoid fibrous histiocytoma of the thigh in a 13-years-old girl is reported. Final diagnosis was made only after surgical excision. A review of the literature with a discussion of the particularity of this case are also examined in order to achieve the best surgical strategy and the lower risk of recurrence.

Key Words:

Histiocytoma, Cistic fibrous histiocytoma, Fibrohistiocytic tumors, Angiomatoid fibrous histiocytoma.

Introduction

Fibrous histiocytoma is a rare soft tissue tumour generally located in derma and less commonly in deep soft tissues. It occurs as a lonely blue nodular lesion in childhood that progressively grows. Some varieties require special attention to discern between benign and malignant forms.

We report a case of uneventfully surgical excision of angiomatoid fibrous histiocytoma of the thigh in a 13-years-old girl. Final histological diagnosis was a very rare variety of "cystic" fibrous hystiocitoma. Lesion appeared highly vascularised, then classified as angiomatoid. Abundant gaps of hemosiderin as a consequence of continue hemorrhagic events inside, developed into a central big cavity that conferred the aspect of "cystic" hystiocitoma.

Case Report

A 13-years-old female patient was referred to our Surgical Unit with a diagnosis of an enlarging mass on the anterior surface of the right thigh. Her mother reported that the lesion firstly appeared at the age of 4 years and by Ultrasound Doppler it was demonstrated to be a 6 mm hypoechogenic nodule with intense vascularisation inside (Figure 1). As the child was completely asymptomatic, no therapy was given and no further investigations were performed. Later in time, parents observed that the lesion progressively enlarged, becoming of a intense blue colour on the surface and changing in size if the girl practiced physical exercise.

A new Ultrasound Doppler showed an increase up to 25 mm (Figure 2) in the maximum diameter of the lesion and the presence of multiple septa covered by a rich plait of collateral vessels. The lesion was also completely plunged in the dermal tissue, but it could be possible to find a surgical plan for the excision by separating it from the vascular branches directed to the below muscles. CT scan of the right inferior leg was subsequently executed: the lesion was confirmed to be a solid oval nodule of 25 × 16 × 26 mm compatible with a cavernous angioma whose refilling was indirectly provided by the femoral artery (Figure 3).

Clinical diagnosis was mistaken as hemangioma. Patient underwent a day surgery intervention, but in general anesthesia, according to the young age and the scarce cooperation of the girl. We proceeded as following described: first we made a rhomboid incision on the skin above the swelling, then we isolated very carefully and excised the lesion, tying the only vessel observed to feed the supposed hemangioma. In the end, we sutured by haemostatic stitches to avoid any risk of subcutaneous bleeding. Patient was discharged a few hours later in lack of complications. On the seventh post operative day she came at visit and stitches were removed.

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Figure 1. Ultrasound Doppler demonstrated a 6 mm hypoechogenic nodule with intense vascularisation inside (age of 4 years).

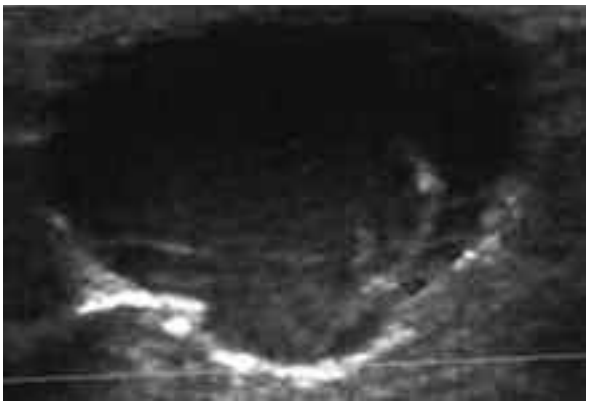

Figure 2. Ultrasound Doppler showed an increase up to 25 mm in the maximum diameter of the lesion and the presence of multiple septa covered by a rich plait of collateral vessels (age of 13 years).

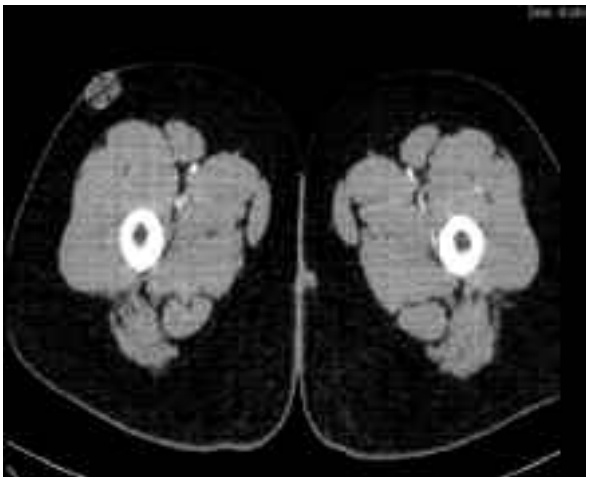

Figure 3. CT scan of the right inferior leg was subsequently executed: the lesion was confirmed to be a solid oval nodule of 25 × 16 × 26 mm compatible with a cavernous angioma whose refilling was indirectly provided by the femoral artery.

On hystologic examination, the entire lesion appeared as a well circumscribed and rounded unilocular cyst located into dermis. Inside there were variably sized hemorrhagic spaces and solid sheets of round to ovoid to short spindly cells surrounded by marked fibrosis and chronic inflammation. Fibroblast-like cells arranged in a storiform pattern. Multiple and diffuse areas of vascular proliferation with massive hemosiderin deposition were present, too. Rare Touton type giant cells were described. Nuclear pleomorphism, mitotic figures and necrosis were absent (Figure 4a, 4b, 4c, 4d).

Finally, diagnosis of "cystic variety" of fibrous histiocytoma, angiomatoid subtype, was made.

Discussion

Fibrohistiocytic tumours are generally located in derma. Classically we distinguish two different forms: benign and malignant ones. Common forms are recognizable easily; some varieties exist yet, like for example the angiomatoid subtype, that is rarely observed and then hardly to characterize. Correct differential diagnosis in the regards of malignant form is of basic importance for the right therapeutic approach[1,2].

Angiomatoid fibrous histiocytoma (AFH) was first described by Enzinger in 1979[3] as an angiomatoid variant of malignant fibrous histiocytoma. Most common localization involves the upper (50%) and inferior arts (20%). AFH was thought to be a relatively aggressive tumor. Among the 24 patients followed up by Enzinger, 11 (46%) experienced local recurrence after excision, 5 (20%) had metastasis and 3 (12%) died of metastasis. Calonje and Fletcher's casuistry in 1995[4] and one year later Zelger et al[5] confirmed the hardly diagnosis and easily misunderstanding of AFH, that were initially interpreted as vascular tumours, melanocytic nevi or dermatofribromas. In particular, Calonje and Fletcher report a series of 40 cases predominantly localized in upper (40%) and lower limbs (20%). In the majority of them, diagnosis was correctly expressed only after surgical excision. Recurrence rate was of 19%.

In 1990 Costa and Weiss[6] affirmed that AFH is a tumor with a low-grade malignant potential. In a total casuistry of 94 cases, local recurrence occurred in 11 patients (12%). All of them underwent a curative re-excision. Five patients devel-

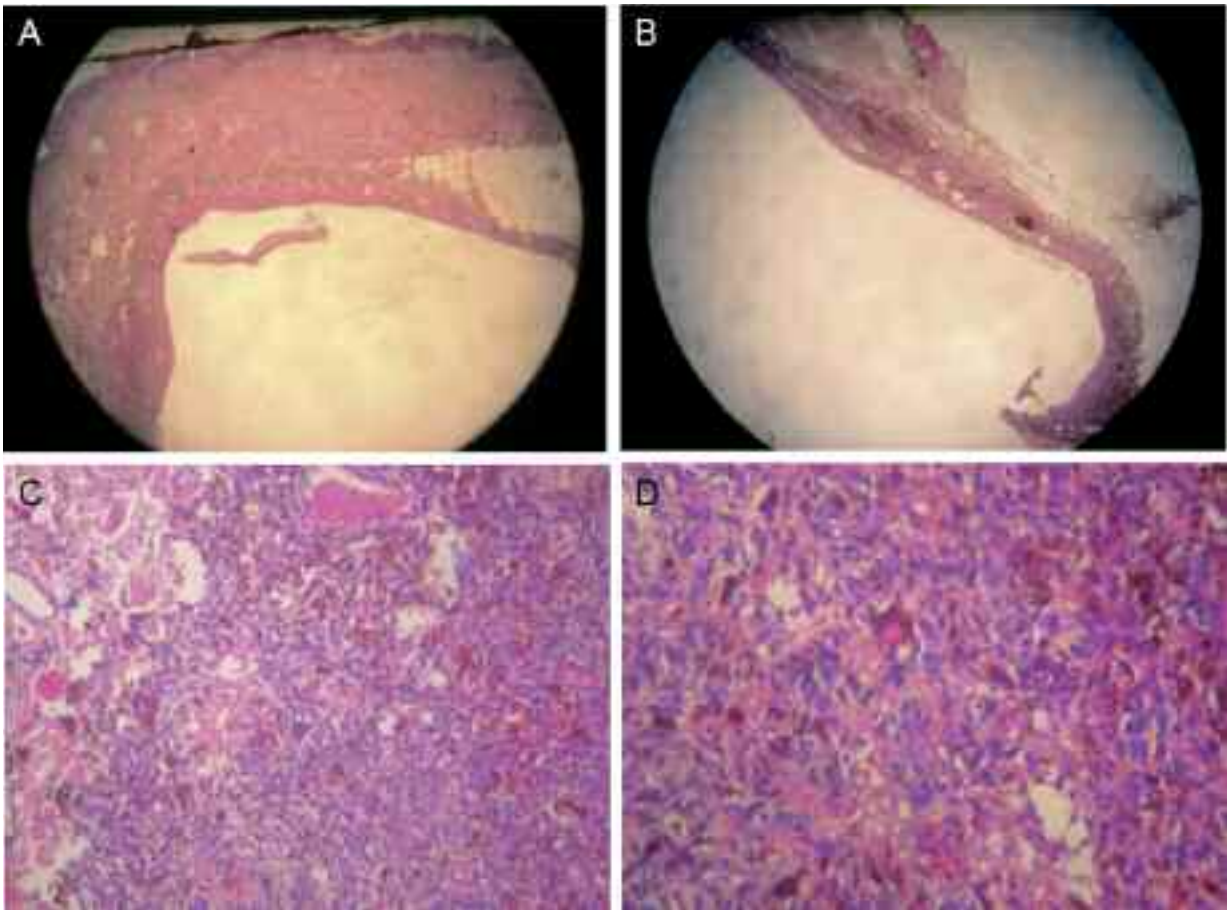

Figure 4. ***A-B,*** Low-magnification (×2) shows a well circumscribed unilocular cystic lesion located into the dermis with partial extension into the superficial subcutaneous fat tissue. ***C,*** Medium magnification (×10) revealed a mainly fibrous lesion composed by fibroblasts arranged in a storiform pattern associated with a striking vascular proliferation, extensive hemorrage and hemosiderin deposition. Hyalinized collagen lines are entrapped by spindle cells at the peripheral border of the lesion. ***D,*** High magnification (×40) shows as the lesion is composed by fibroblasts, foam and histiocyte-like cells phagocyting hemosiderin and occasional Touton type multinucleated giant cells. (Hematoxylin-Eosin).

oped metastasis (5%), but only one (1%) died for distant metastasis.

Another important aspect to consider for differential diagnosis regards flow cytometric analysis. A diploid DNA content has been demonstrated n the majority of AFH cases. This is in contrast with other variants of malignant fibrous histiocytoma (MFH), in which an aneuploid DNA pattern is usually detected[7,8]. On the basis of these observations, World Health Organization Committee for the Classification of Soft Tissue Tumors identifies AFH separately than MFH. In particular the main difference between these two entities concerns low malignant potential of AFH instead of MFH[1].

The present case shows histological aspects of a very rare cystic variety of angiomatoid fibrous histiocytoma. The cavity is formed by a well circumscribed unilocular cystic lesion located in dermis with partial extension to the superficial subcutaneous fat tissue. The fibrous lesion presents fibroblasts arranged in a storiform pattern associated with a striking vascular proliferation, extensive hemorrage and hemosiderin deposition[4,7].

In our report initial diagnosis resulted wrong, because we assumed that the girl was affected by a cavernous angioma on the basis of echotomography and TC scan. The difficulty of an early recognition of angiomatoid fibrous histiocytoma is probably conditioning also the recurrence of the tumour.

Our case is of recent observation, so that we have not a sufficient time of follow up to establish the definitive result. As written before, literature invites to be careful because this kind of lesions is associated predominantly with a high re-

currence incidence, but luckily with a low malignant potential. Surgery excision of the fibrous histiocytoma should be as large as possible, like if we are in front of a malignant lesion, paying attention to regional node involvement, as Calonje and Fletcher signed in a case of recidive fibrous hystiocitoma[4].

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