Aneurysmatic dermatofibroma

Dermatofibroma aneurismático

Review Article

ABSTRACT

A present a clinical case of a rare variant of dermatofibroma (aneurysmatic type), is presented in a 72-year-old female patient, the lesion was located in the right arm's flexural fold, resulting from mesenchymal proliferation associated with blood vessels and tissular hemorrhage, with its own well defined histological characteristics. The objectives are to describe the rarity of the lesion and its histopathologic importance, when compared to other benign and malignant tumors in the differential diagnosis, in addition to the terminology currently used.

Keywords: histiocytoma, benign fibrous; histiocytoma, malignant fibrous; hemosiderin.

RESUMO

Apresenta-se caso clínico de variante rara de dermatofibroma (tipo aneurismático) em paciente do sexo feminino de 72 anos de idade, cuja lesão se localizava na dobra flexural do membro superior direito, resultante de proliferação mesenquimal associada à vasos sanguíneos e hemorragia tecidual, com características histológicas próprias e bem definidas. Os objetivos desta descrição foram a raridade da lesão e a importância que ela assume, do ponto de vista histopatológico, quando comparada no diagnóstico diferencial com outros tumores benignos e malignos e a terminologia utilizada atualmente

Palavras-chave: *histiocitoma fibroso benigno; histiocitoma fibroso maligno; hemossiderina.*

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INTRODUCTION

This case report's objective is to describe a special dermatofibroma sub-type, seldom mentioned in the literature, known as "aneurysmatic dermatofibroma," "angiomatoid fibrous histiocytoma," or hemosiderotic dermatofibroma, which is characterized by its very low incidence (less than 2%) and its importance in the differential diagnosis regarding other types of tumors.[1]

Although its etiology is unknown, several authors have observed that it is correlated to the extravasation of erythrocytes from the vascular walls to the vascular cystic spaces. This process is caused by repeated micro-traumas that lead to the dissection of areas of the tumor, forming typical fissures and causing the phagocytosis of the hemosiderin by the tumorous cells.[2,3]

From a clinical perspective, the morphologic characteristics correspond to a nodular or tumorous lesion (sometimes of cystic consistence) which can be larger than the usual dermatofibromas – ranging from 0.5 to 2 cm in diameter, with a smooth or squamous surface, and colors varying from dark red to brownish or black. It is a benign tumor that originates in the dermis and can reach the subcutaneous layer.[4] It can grow quickly – which can cause pain due to intralesional haemorrhage. These tumors occur more frequently in the lower limbs, however they can appear in other areas, as in the case

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described in this review.

It is more common in female patients older than 30.5 A definitive diagnosis is established by a histopathologic examination characterized by a significant extravasation of erythrocytes and hemosiderin. Since it is frequently confused with vascular or melanocytic tumors such as melanoma, sarcoma of Kaposi, angiosarcoma and angiomatoid fibrous histiocytoma, an exact diagnosis of the lesion is vital for the patient's prognosis. [4,5]

CASE REPORT

A 72-year-old female patient, originally from Sao Paulo, Brazil, presented a single tumor, covered by smooth, violaceous and brilliant skin, of cystic consistency. The tumor was about 2 cm in diameter, located in the right arm's flexural fold, and was associated with the retraction of the surrounding skin (Figure 1). Following a period of slow and gradual growth, it developed quickly in the months prior to diagnosis, which coincided with the onset of pain.

The patient underwent an exeresis of the lesion with 0.5 cm margin and the material was subjected to histopathologic examination (Figure 2). The histological sections revealed evidence of fusocellular dermal proliferation with scarcely defined borders, with a central cavity full of eosinophilic and amorphous material with cholesterol crystal clefts (Figure 3).

The fusocellular proliferation did not show atypias, presenting small-caliber ectasic vessels and hemosiderin deposits in multiple points (Figure 4). With these findings, an aneurysmatic fibrous dermatofibroma diagnosis was established.

DISCUSSION

Gross and Walbach first described the aneurysmatic variant of dermatofibroma in 1943, and discussed its relationship with sclerosant hemangioma. In 1966, Ariston and Reed described lesions consisting of areas typical of dermatofibromas, intermingled with foamy histiocytes, hemosiderin and spaces full of erythrocytes, without endothelial lining. It was not until 1981, however, that Santa Cruz and Kyriakos denominated three "aneurysmatic fibrous histiocitomas" out of a series of 17 cases.6 The terms "aneurysmatic" or "angiomatoid" describe a histological variant of dermatofibromas within which there are vascular spaces that dissect the stroma of the tumor, forming true venous lakes in some cases. Due to the fact that those vascular spaces are not lined by endothelium and are not abnormal dilatations of the vascular system, some authors believe that the denomination "aneurysmatic" is incorrect.6,7 Likewise, it would not be correct to refer to it as "angiomatoid" since there is no proliferation of the vascular component, and consider it more appropriate to use the term "hemorrhagic" once there is a predominance of the extravasation of erythrocytes into the dermatofibroma cells. [7]

Histologically, dermatofibroma presents numerous variants described as cellular, epithelioid, aneurysmatic, hemangiopericytoid, atrophic, fibrocollagenous, and pseudosarcomatous.

Such denominations are all characterized by the more important histological finding, that typical dermatofibroma features are found in a large part of the lesions. However, it is worth noting that those subtypes do not have clinical relevance, but histopathologic importance since many times they are confused with malignant lesions.[8]

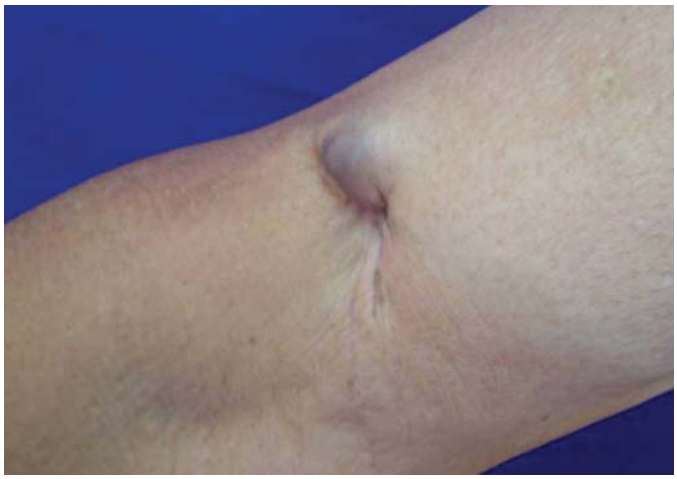

Figura 1- Cystic consistence lesion located in the flexural fold of the right arm

The diagnosis is established by the histopathological findings. The neoformation located in the dermis is composed of myofibroblast, fusiform cells and histiocytes containing hemosiderin in its fibrous stroma, Touton giant multinucleated cells and blood-filled spaces without endothelial lining, occupying most of the lesion. The findings vary from narrow clefts to large cavernous cysts. In addition, points of hemorrhage of the stroma adjacent to the vascular channels and extravasated erythrocytes among the cells of the tumor were observed, with the presence of solid areas with a dermatofibromatous appearance. Hyperplasia and irregular acanthosis are also observed in the epidermis.[8]

Currently it is accepted that the fusiform cells of the cutaneous fibrohistiocytomas originate from dermal dendrocytes intermixed with fibroblasts and myofibroblasts, forming variable amounts of collagen, accompanied by

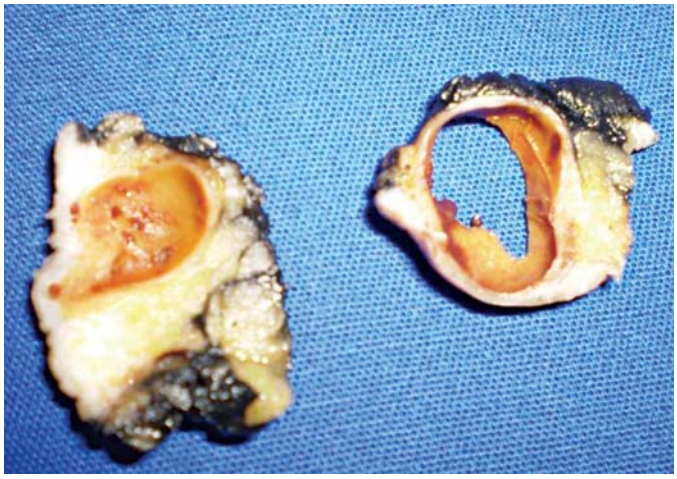

Figura 2 - Macroscopic appearance of the lesion's cystic cavities

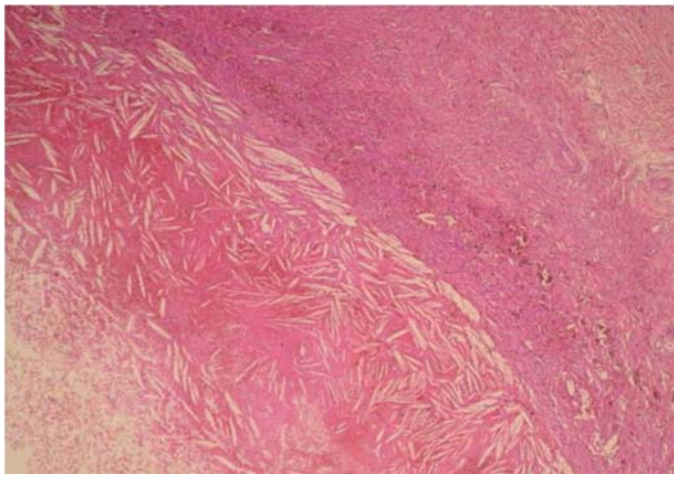

Figura 3 - Dermal fusocellular proliferation froth with eosynofilic and amorphous material, andclefts of cholesterol crystal

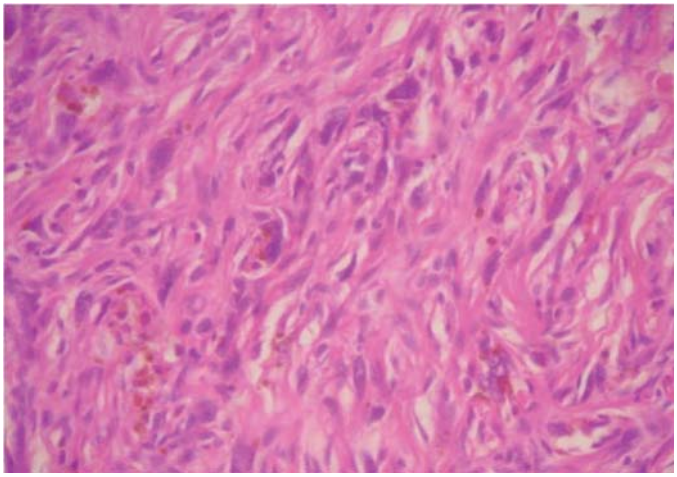

Figura 4 - Fusocellular proliferation without atypias, with small-caliber ecstasic vessels and hemosiderin deposits

histiocytes in different stages of maturation.[8,9] Santa

Cruz and Kyriakos proposed that the aneurysmatic variant can originate inside a fibrohistiocytoma or common dermatofibroma, when small amounts of erythrocytes extravasate from the capillary vessels within the tumor. Therefore, the older erythrocytes that converted into hemosiderin are phagocyted by the histiocytes with the fibro-histiocytoma called "hemosiderotic" in that phase.9 With the continuous extravasation of erythrocytes, cracks or clefts appear, which entails a loss of stromal support and results in an increase in the tumor's internal pressure, leading to the dilatation that characterizes the typical "angiomatoid" appearance. In this phase the aneurysmatic dermatofibromas appear (the hemosiderotic type would be the initial phase of the aneurysmatic type).[9] The etiology of these aneurysmatic or angiomatoid cavities is not clear. Some authors maintain that this phenomenon could happen in areas of hypocellularity or secondary to repeated traumas or micro-traumas that would trigger micro-hemorrhages, unchaining the string of events already described.[9]

From the immunohistochemical perspective, the fusiform cells of the dermatofibroma are reactive to factor XIIIa (in stages in which hemosiderin deposits are low), demonstrating the participation of dermal dendrocytes. Mac 387 (histiocytotic antigen), vimentin, smooth muscles' actin, and CD57 reveal fibroblastic and myofibroblastic differentiations, and are negative for factor VIII, desmin and S-100 proteins.10 Ultrastructurally, these lesions present histiocytes and fibroblasts containing few lysosomes, and lipids with moderately developed endoplasmic reticulum. They also present endothelial cells with an absence of morphologic abnormalities and without a rupture of the basal lamina. The differential diagnosis is established, fundamentally, between benign vascular lesions – especially hemosiderotic hemangioma, and with some malignant vascular lesions such as the malignant fibrous angiomatoid histiocytoma (which occurs in subcutaneous cellular tissue, muscle and periosteum) – and melanoma, which has very different histological aspects. Other differential clinical diagnoses include the papular, nodular or plaque-shaped lesions of Kaposi's sarcoma (presenting immunoreactivity to CD4 and an absence of fibro-histiocytic cells) and angiosarcoma (atypical endothelial cells with separate of collagen sheafs are observed).[10]

Selecting the appropriate treatment involves surgical resection, with an advised margin of at least 3 to 5 mm. According to some studies, the rate of recurrence was about 20%, meaning that the patients' clinical follow up is necessary.[9,10] ●

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