Aneurysmatic dermatofibroma

Dermatofibroma aneurismático

ABSTRACT

A present a clinical case of a rare variant of dermatofibroma (aneurysmatic type), 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 a 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; hemosiderina.

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.

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.

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. 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...
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.5,6

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
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
ttrue 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"
when 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.5

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
erthrocytes 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.5

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
histiocytes in different stages of maturation.\textsuperscript{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 fibrohistiocytoma called “hemosiderotic” in that phase.\textsuperscript{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).\textsuperscript{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.\textsuperscript{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 (histiocytic antigen), vimentin, smooth muscles’ actin, and CD57 reveal fibroblastic and myofibroblastic differentiations, and are negative for factor VIII, desmin and S-100 proteins.\textsuperscript{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 popular, 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).\textsuperscript{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.\textsuperscript{10,11}

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