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Aneurysmal dermatofibroma with exuberant presentation

Dermatofibroma aneurismático com apresentação exuberante

Case report

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ABSTRACT
Aneurysmal dermatofibroma (ADF) is a rare benign tumor originating in the dermis. It is most common in the lower limbs of women over 30 years. Its etiology is unknown, and histopathological examination confirms the diagnosis. This paper presents a case of ADF with an exuberant presentation in a young female patient with a pedunculated tumor in the right pretibial region, measuring 12 cm in greatest diameter. This report aims to demonstrate an infrequent variant of dermatofibroma with significant growth and emphasize the importance of lesion excision and follow-up because of the high recurrence rate.
Keywords: Histiocytoma, Benign fibrous; Dermatology; Dermoscopy

RESUMO
O dermatofibroma aneurismático (DFA) é um tumor benigno raro, originado na derme. É mais frequente em membros inferiores de mulheres acima de 30 anos. Sua etiologia é desconhecida, e o exame histopatológico confirma o diagnóstico. Este trabalho apresenta um caso de DFA com apresentação exuberante, em paciente jovem, do sexo feminino, com tumoração pedunculada em região pré-tibial à direita, medindo 12cm no maior diâmetro. O objetivo deste relato é demonstrar uma variante pouco frequente de dermatofibroma com crescimento importante e ressaltar a importância da exérese da lesão e seguimento pela alta taxa de recorrência.
Palavras-chave: *Histiocitoma fibroso benigno; Dermatologia; Dermoscopia*

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INTRODUCTION

Dermatofibromas (DF) are common benign tumors originating in the dermis and may extend to the subcutaneous tissue. They are more frequent in the trunk and extremities of young adults, with a predominance in women from the third decade of life.[1]

Considering clinical and histopathological characteristics, DF is classified into multiple variants that include aneurysmal, hemosiderotic, cellular, epithelioid, atypical, lipidized, clear cell, palisading, atrophic, keloidal, granular cell, myxoid, lichenoid, among others. Thus, the diagnosis can be challenging.[2,3]

Aneurysmal dermatofibroma, also known as aneurysmal benign fibrous histiocytoma, is an uncommon form representing 1% to 7% of cases.2 Santa Cruz et al. first described it as an "aneurysmal fibrous histiocytoma".[4] It is a rare variant of cutaneous fibrous histiocytoma that results from the proliferation of blood vessels and hemorrhage in a fibrous histiocytoma.[5] It has an unknown etiology. Several authors have noted a correlation with the extravasation of red blood cells in the vascular wall due to repeated microtraumas that dissect the tumor area forming fissures and resulting in hemosiderin phagocytosis by tumor cells.[1]

CASE REPORT

A 24-year-old woman, skin phototype III, was referred from primary health care because of a tumor in the right leg due to a history of a papule in the right distal pretibial region for ten years. The lesion was asymptomatic, with progressive growth, mainly after a twin pregnancy in 2019.

Physical examination revealed a pedunculated tumor in the right pretibial region, measuring 10x12 cm (Figure 1), with a positive transillumination test (Figure 2).

Dermoscopy showed white linear structures and a homogeneous background ranging from blue-purplish to red-brown (Figure 3).

An excisional biopsy was performed with primary closure of the surgical wound.

The anatomopathological study revealed a well-defined benign neoplasm with expansive borders, characterized by fibrohistiocytic proliferation without atypia, exhibiting collagen entrapment in the periphery and dilated and congested capillaries, with extravasation of red blood cells and numerous hemosiderophages, accompanied by the formation of aneurysmal pseudovascular spaces with fibrinohematic material (Figures 4 and 5). Fluid drain cytology was negative for neoplastic cells.

DISCUSSION AND CONCLUSION

DF usually presents clinically as a papule, nodule, or erythematous brownish or violaceous tumor. The aneurysmal type is a rare variant occurring in less than 2% of DF. It is larger than the classic DF, and it can be painful due to intralesional hemor-

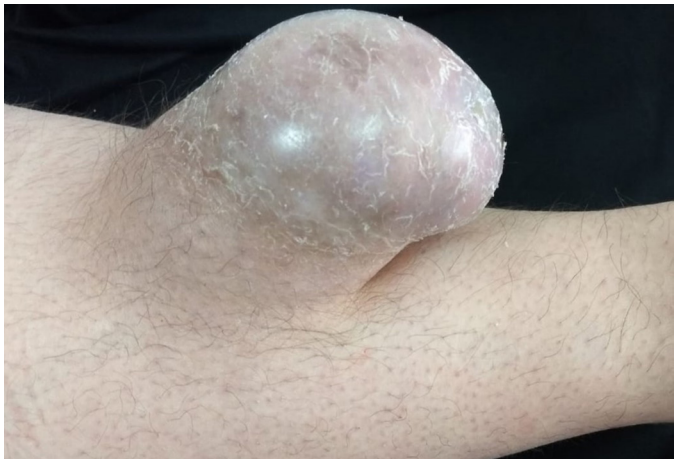

Figure 1: Pedunculated tumor in the right pre-tibial region, measuring 10x12cm

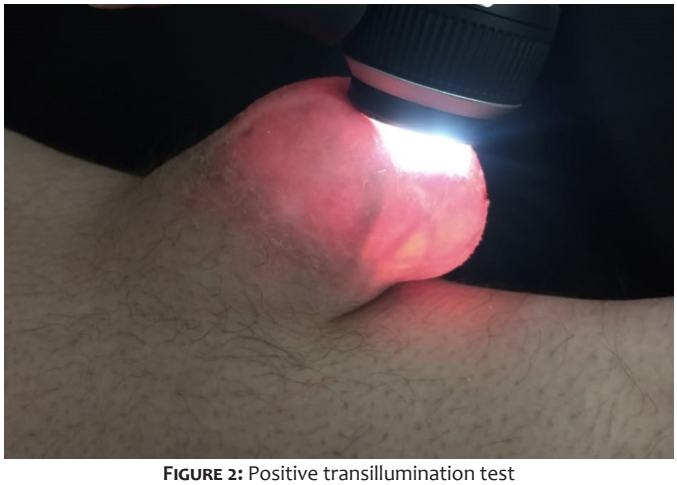

Figure 2: Positive transillumination test

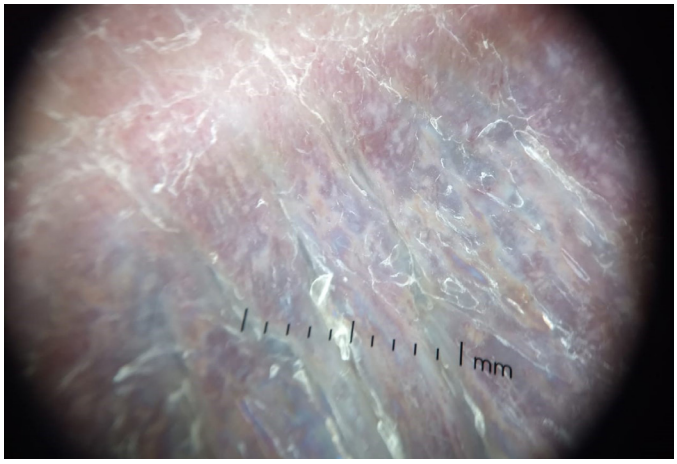

Figure 3: Dermoscopy with white linear structures and homogeneous background ranging from purplish-blue to brownish-red

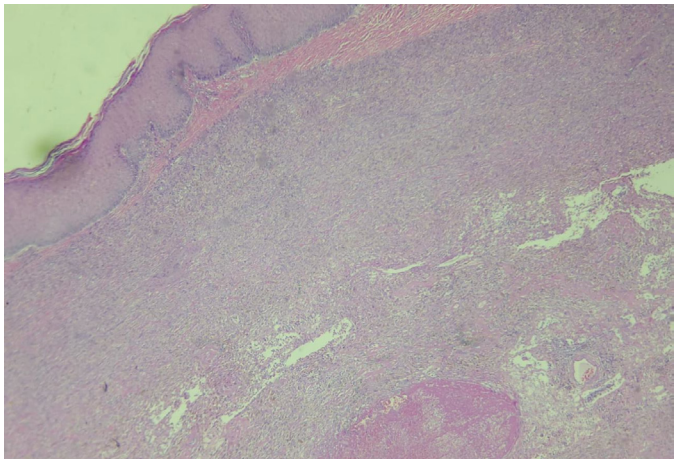

Figure 4: Anatomopathological examination at 4x magnification showing a well-delimited and expansive lesion composed of spindle cells, and aneurysmal pseudovascular space (H&E staining)

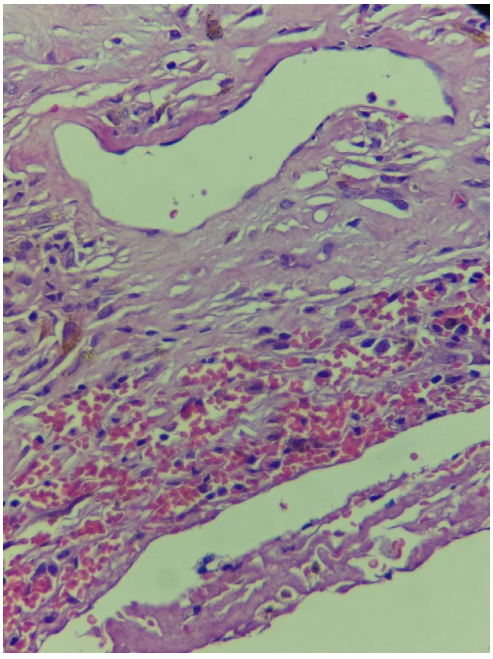

Figure 5: Anatomopathological examination at 40x magnification showing capillary dilation, associated with extravasation of red blood cells, some hemosiderophages and cellular component of fibroblasts and histiocytes (fibrohistiocytic). (H&E stain)

rhages, causing discomfort to the patient and reduced quality of life.[1,6,7] Studies show a predilection for lower limbs (55%), followed by upper limbs (17%), trunk (12%), and head and neck (4%).[8]

Dermoscopy can find known DF patterns. However, the characteristics that suggest aneurysmal DF are white linear structures, brown or purplish background, vascular structures, and a delicate pigmented network in the periphery.[9]

Aneurysmal dermatofibroma is challenging to diagnose due to its rarity, morphological heterogeneity, inconsistent immunological profile, and overlap with a wide spectrum of neoplastic and non-neoplastic, benign, and malignant entities.[10] Its incidence may be underestimated due to overlapping histopathological findings.[11]

Histopathological examination confirms the diagnosis showing a dermal-based cellular nodule composed of intertwined fascicles of thin cells with peripheral collagen entrapment, spaces similar to central hemorrhagic cysts without endothelial lining, extracellular hemorrhage, and hemosiderin-laden macrophages.[12]

Regarding immunohistochemistry, spindle cells of dermatofibroma usually express factor XIII and are negative for immunostaining with S100 HMD45 and CD34.[1,9]

Imaging findings are nonspecific regarding its histogenesis. When originating from soft tissue, X-rays and CT scans show a heterogeneous mass, possibly suggesting cystic and enhancement components. However, they are ultimately inferior to MRI soft tissue detail.[13]

The differential diagnosis is made with hemosiderodic hemangioma, angiomatoid malignant fibrous histiocytoma, and melanoma, whose histological aspects are completely different. Other clinical differential diagnoses include papular, nodular, or plaque-like lesions of Kaposi sarcoma, angiosarcoma, and epithelioid fibrous histiocytoma.[9,14]

The treatment must be conducted with exeresis to differentiate it from malignant lesions. Surgical margins between three and 5 mm are recommended.[1]

The disease prognosis is favorable. Local recurrence after excision ranges from 2% to 10%, and the chance of metastasis is less than 1%.[15,16]

Therefore, the exeresis of the lesions is indicated for diagnostic clarification, and the patient follow-up must be conducted frequently due to the possibility of recurrence. ●

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