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## An aneurysmal histiocytofibroma of the forearm: About a case

Oukarfi Sara<sup>1</sup>, Tahiri Laila<sup>2</sup>, Baybay Hanane<sup>3</sup>, Rasso Asmae<sup>4</sup>, Elloudi Sara<sup>5</sup>, Douhi Zakia<sup>6</sup>, ELfatemi Hind<sup>7</sup>, Mernissi Fatima Zahra<sup>8</sup>

<sup>1, 3-6, 8</sup> Department of Dermatology and Venereology Hassan II University Hospital, Fez, Morocco

<sup>2,7</sup> Anatomy and pathology laboratory, Hassan II University Hospital, Fez, Morocco

#### Abstract

Benign fibrous histiocytoma is a benign tumor with multiple clinical and histological variants. Clinical diagnosis becomes difficult in the face of atypical variants, histology is the key to diagnosis. We report a case of aneurysmal histiocytofibroma of the forearm.

Keywords: Dermatofiroma, aneurysmal Histiocytofibroma, dermoscopy, histology

### Introduction

One of the most common skin tumors is dermatofibroma (DF) or benign fibrous histiocytoma; it represents around 3% of skin biopsy samples received in dermatology laboratories.

DF is easy to diagnose clinically, but diagnosis becomes difficult in variants and atypical cases.

Keywords: Aneurysmal histiocytofibroma, dermatofibroma, dermoscopy, histology.

### Observation

Mrs. B.H, 35 years old, with no particular history, reports for 20 years the appearance of a lenticular lesion of the right forearm having gradually increased in asymptomatic size, becoming ulcerated and bleeding after manipulation, the examination objectified a purple, round, well-defined tumor with regular contours measuring 4 cm along the long axis, of firm consistency, fixed relative to the deep plane, ulcerated in the center with a homogeneous peripheral pigmentation [Figure 1]. In dermoscopy, we visualized a homogeneous peripheral pigmentation, a white ring surrounding the central ulceration, haemorrhagic suffusions and some point vessels. [Figure 2]

Darier ferrand dermatofibrosarcoma, histiocytofibroma, achromic melanoma or epidermoid carcioma have been suspected.

A histological and immunohistochemical study has objectified skin tissue lined with acanthosic epidermis. The dermis is the site of a spindle cell tumor proliferation, of storiform architecture largely dissociated by non-endothelialized vascular clefts. The tumor cells did not present cytonuclear atypia, there was no mitosis seen, in immunohistochemistry, the tumor cells express vimentin and focus CD68 and AML [figure 3, 4, 5, 6]. A panel of markers carried out eliminated vascular tumors, a dermatofibrosarcoma of Darrier ferrand, a melanoma, a carcinoma or a tumor of nervous origin.

We corrected our diagnosis using histology and retained the diagnosis of aneurysmal histyocitofibroma.

The patient was referred to the trauma surgeon for complete excision.



Fig 1: clinical image of BAFH of the forearm



**Fig 2:** Dermoscopic images of BAFH: homogeneous peripheral pigmentation, a white ring surrounding the central ulceration, haemorrhagic suffusions and some point vessels.



Fig 3: Histological image of aneurysmal histiocytofibroma



Fig 4: AML and CD68 staining: shows focal expression



Fig 5: Vimentin staining: shows a spread expression.

## Discussion

Benign aneurysmal fibrous Histiocytomas (BAFH) are rare variants of Dermatofibromas which represent 1.7% of all Dermatofibromas<sup>[2]</sup>.

Described for the first time in 1981 <sup>[1]</sup>, Dermatofibromas are brown or blue solitary nodules or tumors generally measuring 0.5 cm to 2 cm and less than 3 cm <sup>[3]</sup>. BAFHs are generally larger, ranging from 0.5 cm to 4 cm <sup>[2]</sup>. The term "giant Dermatofibroma" has been used for lesions measuring 5 cm or more <sup>[4]</sup>.

The majority of BAFH occurs on the extremities, especially the lower extremities <sup>[2]</sup>. They are also found in the trunk or head and neck <sup>[2]</sup>. They can occur at any age, with a slight predilection for the middle ages <sup>[2]</sup>. There does not appear to be a difference in gender or race.

Common differential diagnoses include melanoma, kaposi's disease, shwanoma, and neurofibroma  $^{[1, 2]}$ .

Several dermoscopic models of DF are described, the most common of which is the central white area in the form of a white scar with a pigmented network at the periphery. <sup>[5]</sup>.

A recent study <sup>[5]</sup> revealed that the discrete peripheral network and the white star-like appearance, the white ring around an ulceration was strongly linked to aneurysmal DF. This can be a very useful sign for differentiating aneurysmal DF and achromic melanoma, especially because this variant of DF is also characterized by polymorphic vessels and linear irregular vessels, which can be confusing and misdiagnosed as a malignant tumor. Other non-specific signs reported are the bluish or reddish central homogeneous zones with white structures and a network of peripheral pigments with vascular structures of varying degrees, this form of ring could be of great help in the diagnosis of cases. Difficult aneurysmal DF.

Histologically are well circumscribed lesions present in the dermis or subcutaneous tissues, there are spaces filled with blood inside the lesion. Any accidental presence of hemosiderin must be accompanied by these spaces filled with blood; the diagnosis cannot be established on the basis of hemosiderin alone <sup>[2]</sup>. These spaces are not vascular in nature, so there is no endothelial wall. However, small blood vessels may be present in the lesion <sup>[2]</sup>. Epidermal hyperplasia, sclerotic collagen and an infiltrate composed of lymphocytes and macrophages are frequently observed <sup>[6]</sup>.

Immunohistochemical stains may be requested to aid in diagnosis; however, there is no specific marker for ABFH. Vimentin is positive, but it is not specific. CD31 and CD34 are negative <sup>[2]</sup>. This benign entity must be differentiated from an angiosarcoma, which is CD31 and CD34 positive.

Another differential diagnosis is angiomatoid (malignant) fibrous histiocytoma (AMFH), which has a distinct multinodular pattern, a thick pseudocapsule and a peripheral inflammatory infiltrate of lymphocytes and plasma cells <sup>[6]</sup>. These lesions are usually found in deep subcutaneous tissue or muscle <sup>[7]</sup>.

Another similar-looking vascular tumor is kaposi disease, which presents a similar type of extravasation of red blood cells, but fibrocytes and macrophages will not be seen in AFH <sup>[7, 16]</sup>. This endothelial tumor is positive for CD31 and CD34 <sup>[8]</sup>, as well as HHV8 <sup>[9, 10]</sup>.

Although considered a benign lesion, the local recurrence rate is higher after surgical removal of the lesion due to diffuse infiltration and the involvement of deeper structures.

### Conclusion

DFs are characterized by a variety of clinical and histopathological findings. The characterization of these variants is important for differential diagnosis and prognosis because they have different probabilities of local recurrence and, in rare and controversial cases, metastases. For this reason, it is important to improve other non-invasive diagnostic tools, including dermoscopy.

### **Conflicts of Interest**

The authors do not declare any conflict of interest.

### Author contributions

All the authors contributed to the writing of this article. The authors also declare having read and approved the final version.

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