

# Aneurysmal Dermatofibroma: Uncommon Presentation of Common Disease

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## Abstract

**Aneurysmal** dermatofibroma is an uncommon, benign variant of dermatofibroma due to the slow extravasation of blood. Clinically, it is difficult to differentiate from other vascular occurring lesions like verrucous hemangioma and malignancies like malignant melanoma and nodular basal cell carcinoma. Hence, an excisional biopsy followed by a thorough histopathological examination is necessary to avoid misdiagnosis. Here, we report a case of a 38 year old female who presented with a brown colored nodular lesion on her left upper back for the last one year, and histopathological examination was consistent with aneurysmal dermatofibroma.

**Key words** : Aneurysmal dermatofibroma; Basal cell carcinoma; Malignant melanoma; Verrucous hemangioma

## Introduction

Dermatofibroma (DF), /benign fibrous histiocytoma, is a commonly occurring cutaneous entity usually centered within the dermis. It is a benign dermal and superficial subcutaneous myofibroblastic proliferation, microscopically mimicking fibromatosis. Dermatofibroma is generally reported on the trunk or extremities in adults between 20 and 49 years old, with women being more frequently affected than men.<sup>1</sup> Based on specific histopathological features, many variants of dermatofibroma have been identified in the literature which include cellular DF, aneurysmal DF, atypical DF, epithelioid DF, atrophic DF, ulcerated DF, erosive DF, and lichenoid DF.<sup>2</sup>

Clinically, they present as dome shaped, firm papules or nodules that usually range from a few millimeters to occasionally up to 2 cm in size.<sup>3</sup> It may show a dimple sign that indicates tethering of the overlying epidermis to the underlying lesion.<sup>2</sup> Aneurysmal dermatofibroma is one of the uncommon histological variants of dermatofibroma, which closely mimics vascular lesions. It is usually larger than typical dermatofibroma and may show rapid growth.<sup>4</sup> Here we report a rare and interesting case of aneurysmal dermatofibroma which resembled verrucous hemangioma clinically but

histopathological examination helped us in clinching the diagnosis.

## Case Report

A 35 year old female presented with an asymptomatic single brown colored nodular lesion since one year on her left upper back, which was slowly increasing in size. The patient had no prior history of trauma at the same site. Physical examination revealed a single, 1\*1 cm size, well defined, nontender, erythematous nodule, having a violaceous hue with central reddish brown crusting and irregular puckered margins present over the left side of the upper back. (Figure 1)

Dermatoscopy (10x) of the lesion showed a central bluish homogenous area with overlying whitish structures and a peripheral delicate pigment network along with vascular structure (Figure 2).

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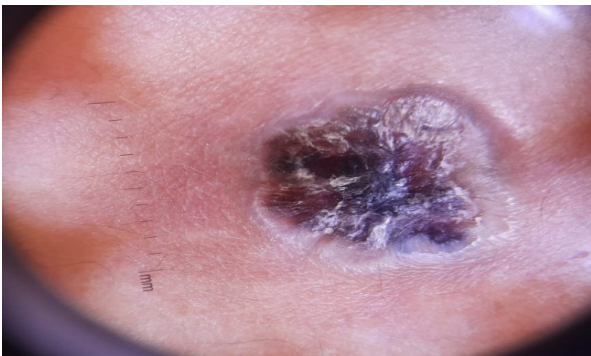
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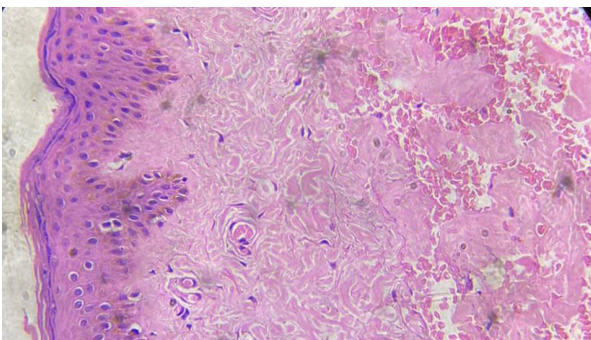
**Figure-1:** single ,1\*1 cm size well defined, non tender, erythematous nodule, having a violaceous hue with central reddish brown crusting and irregular puckered margins present over the left side of the upper back.



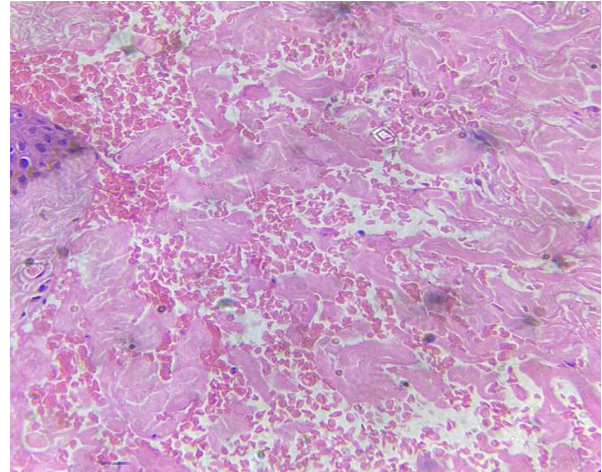
**Figure-2:** On dermatoscopy (10x) = central bluish homogenous area with overlying whitish structures and a peripheral delicate pigment network along with vascular structure

Based on clinical evaluation, provisional diagnoses of verrucous hemangioma, nodular basal cell carcinoma, and aneurysmal dermatofibroma were kept, and the patient was evaluated further.

Ultrasonography of the lesion showed an approximately 9 x 5mm sized, well defined hypoechoic lesion noted in the subcutaneous plane with evidence of internal vascularity (possibility of slow flow venous malformation or melanocytic nevus). The lesion was then completely excised and sent for histopathological examination. H&E stained section of a skin biopsy revealed hyperplastic squamous epithelium with an underlying dermal lesion composed of short spindle and ovoid cells with irregularly shaped nuclei arranged in a storiform pattern along with vascular proliferation (Figure 3,4). Based on the clinical presentation and characteristic histopathological findings, the lesion was finally diagnosed as an aneurysmal dermatofibroma.



**Figure-3:** 10x magnification of a haematoxylin and eosin stained section of skin biopsy revealed hyperplastic squamous epithelium with an underlying dermal lesion composed of short spindle and ovoid cells with irregularly shaped nuclei arranged in a storiform pattern along with vascular proliferation.



**Figure-4:** Vascular proliferation seen at 20X magnification

### Discussion

Aneurysmal dermatofibroma is considered to be a rare variant of dermatofibroma. It was first described by Santa Cruz and Kyriakos in 1981.<sup>4</sup> Clinically, aneurysmal dermatofibromas are characterized by benign, slow-growing, smooth nodular lesions that are cystic in consistency and may be associated with pain. They vary in color, ranging from black, blue, and dark red in the early stages to brown and yellow in the later stages. The color is because of extravasation of erythrocytes and the presence of hemosiderin, and variations in color are likely due to various expressions of red blood cell degradation.<sup>1</sup> Aneurysmal DF is reportedly associated with a vascular tumor-like pattern or melanoma-like pattern on dermoscopy.<sup>5</sup>

Histopathologically, aneurysmal DF shows large blood filled spaces bound by densely aggregated siderophages, but without the typical endothelial lining of vascular structures. Extravasation of red blood cells is seen along with sheets of large histiocytes and siderophages which are interspersed with small capillaries without endothelial atypia.<sup>3</sup>

Based on histological findings, features of aneurysmal DF can be confused with vascular or malignant tumors; hence, expert evaluation is necessary in view of the rarity of the occurrence of this disease. Additional investigations that help in differentiation include special stains like Prussian blue and immunohistochemistry (IHC). Siderophages in aneurysmal DF are Prussian blue positive, and IHC is positive for vimentin and negative for factors CD31, CD34, desmin, CD8, and XIIIa.<sup>4</sup> In our case, the lesion closely mimicked verrucous hemangioma and pigmented nodular basal cell carcinoma on clinical examination as

well as dermatoscopy. but histopathology showed characteristic findings that suggested aneurysmal dermatofibroma. Due to resource constraints, we could not do IHC or special stains.

### Conclusion

Aneurysmal dermatofibroma is difficult to diagnose when it presents with atypical morphological features.

So this case report re-emphasizes the need for complete excision followed by a histopathological examination in all nodular lesions where the list for differential diagnosis is long and clinical as well as dermoscopic features are overlapping.

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