A Solitary Firm Nodule Over Back

Dear Editor,

An adolescent male presented with a four-year duration of a single asymptomatic swelling over the right upper back. The swelling was not associated with any discharge. The patient could not relate the swelling to a prior history of trauma. He received multiple intralesional triamcinolone acetonide injections without any significant improvement. Cutaneous examination revealed a solitary dome-shaped, smooth, reddish-brownish nodule of size 2.0 × 1.0 cm on the right upper back with a small focus of atrophy. The swelling was firm to hard on palpation with few intervening areas of soft consistency [Figure 1a]. The rest of the cutaneous and systemic examination was normal. Dermoscopy of the nodule showed a central multicolored pattern surrounded by an area of radial white streaks, which was in turn surrounded by a gray-brown structureless area. Focal hairpin vessels were also noticed [Figure 1b]. Histopathology revealed acanthotic and pigmented epidermis and ill-defined unencapsulated spindle cell proliferation with a central pseudovascular space, without any endothelial cell lining surrounded by siderophages [Figure 2a and b]. Focal areas of collagen trapping were also seen in the periphery of the tumor [Supplementary Figure 1]. Other features like epidermal acanthosis and Touton giant cells were not seen. Immunohistochemistry was negative for S100 and CD34. A diagnosis of aneurysmal dermatofibroma (ADF) was made and the lesion was excised surgically without any recurrence even after five months.

ADF, a rare variant of dermatofibroma representing 1.7% of all cases, presents commonly in middle age as a solitary nodular swelling with cystic consistency.[1] The usual sites of involvement are extremities, but the trunk can also be involved. It may be associated with rapid growth, pain, and higher recurrence rate of around 20% following surgical excision, and can be mistaken as a malignancy such as malignant melanoma and Kaposi's sarcoma.[2] The vascular component results from sudden intratumoral hemorrhage and the formation of pseudovascular space. The same is responsible for the soft consistency of the well-known firm nodule of ADF, its rapid increase in size and a relatively larger size of ADF as compared to dermatofibroma. Dermoscopic features of ADF described previously include multicolored rainbow patterns representing underlying vascular nature, pigment network, blue homogenous area, shiny-white structures, branched streaks and vascular structures which include dotted, polymorphic and linear irregular vessels.[1,3] Multicolored pattern and gray-brown structureless areas seen on dermoscopy in our case are consistent with previous studies. Additional findings seen

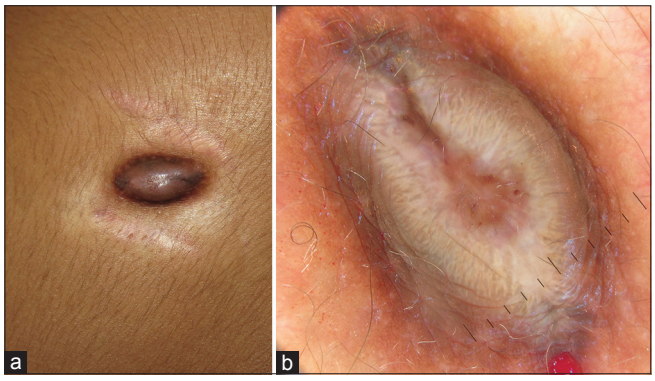

Figure 1: (a) Solitary dome-shaped reddish-brown nodule with peripheral brown pigmentation present on the upper back. (b) Dermoscopy showing a central multicolored pattern surrounded by an area of radial white streaks, and a gray-brown structureless area in the periphery along with focal hairpin vessels (Heine Delta30 dermoscope 10×)

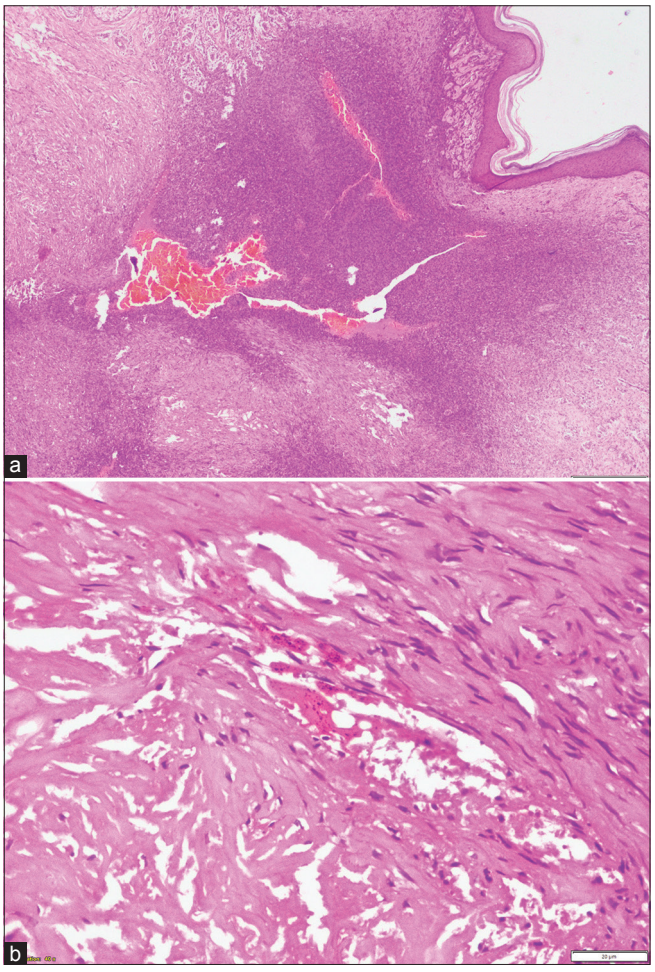

Figure 2: (a) Baseket-weave orthokeratosis and pigmented epidermis. Dermis contains ill-defined unencapsulated proliferation of spindle cells with multiple pseudovascular spaces (H and E, 40×). (b) At higher magnification, vascular space without lining endothelium is surrounded by spindle-shaped cells and siderophages (H and E, 400×)

in our case were radial white streaks and focal hair-pin vessels. A multicolored pattern under the dermoscopy has been described before, as in our case and represents the underlying vascular nature of the nodule. On histopathology, ADF shows short spindle cells and histiocyte-like cells, often in a storiform pattern, intermingled with a variable number of hemosiderin-containing giant cells and macrophages. Blood-filled spaces without lining endothelium are seen within the tumor.[4] Dermatofibroma shows CD34 negativity and Factor XIIIa positivity on immunohistochemistry.

Our patient presented with a solitary asymptomatic firm nodule over the upper back. The clinical possibilities for such a presentation include keloid, dermatofibroma and lobomycosis. Keloid can be ruled out in the absence of a history of trauma before the onset of lesions, with no other lesion to suggest keloidal tendency and non-responsiveness to intralesional corticosteroid injection. Lobomycosis was excluded because of the non-endemicity of the area and solitary lesion over nonexposed sites. Solitary pigmented nodule and firm consistency favored the diagnosis of dermatofibroma. In addition, ADF can be considered due to the intervening soft areas caused by intratumoral hemorrhage.[1] A dermoscopy of the keloid will show vascular structures that were not present in our case. The dermoscopy of lobomycosis is not well described. Radial white streaks and a central white area can be consistent with the dermatofibroma.[1] Keloid and lobomycosis do not present diagnostic problems in histopathology and can be easily differentiated from ADF. However, ADF can be confused in histopathology with angiomatoid fibrous histiocytoma and vascular tumors like spindle cell hemangioendothelioma, Kaposi sarcoma or angiosarcoma.[2,4] Angiomatoid fibrous histiocytoma is characterized by the presence of monomorphic, rounded, eosinophilic, desmin-positive cells, surrounded by prominent lympho-histiocytic and plasma cell infiltrate.[2,4,5] Spindle cell hemangioendothelioma is composed of true cavernous vascular spaces, papillary intraluminal structures, and solid spindle cell areas with focally vacuolated cells.[6] Kaposi sarcoma shows the presence of CD 34 positive, spindle cells forming slit-like spaces containing red blood cells. Angiosarcoma is characterized by anastomosing vascular channels lined by plump, pleomorphic, rounded, polygonal, or spindle-shaped endothelial cells which show CD34, and Factor VIII positivity.

To conclude, aneurysmal dermatofibroma requires a high degree of suspicion for diagnosis. This is especially when a keloid-like lesion is present without any history of prior trauma on sites unusual for keloid formation and no response to usual keloid treatments. The presence of soft consistency in such a lesion will point toward aneurysmal DF and will avoid unnecessary intralesional corticosteroid injections and their side effects.

Acknowledgment

We thank the patient for granting permission for clinical photography.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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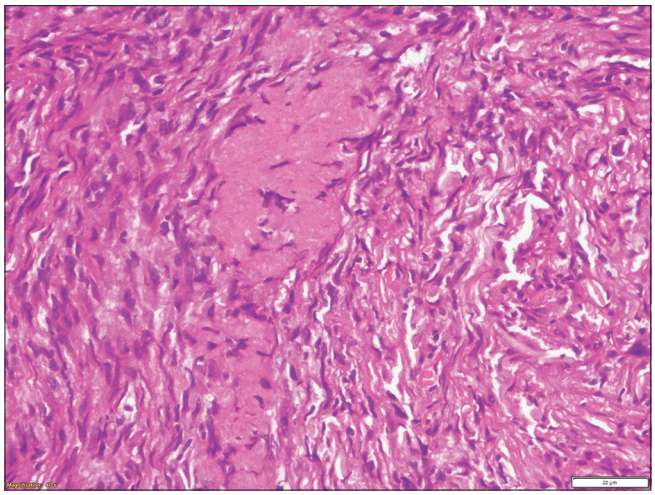
Website: https://journals.lww.com/idoj

Quick Response Code

DOI: 10.4103/idoj.idoj_425_23

How to cite this article: Gupta P, Garg S, Sethy M, Behera B, Thakur V. A solitary firm nodule over back. Indian Dermatol Online J 2024;15:652-4.

Received: 02-Jun-2023. **Revised:** 27-Oct-2023.
Accepted: 06-Nov-2023. **Published:** 20-May-2024.



Supplementary Figure 1: Histopathological image showing collagen trapping in the periphery of the lesion (H and E, 400×)