A Dusky Red Plaque with Satellite Lesions

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A 27-year-old man presented with a dark colored lesion on the left upper back since 6 years. It began as a small, dark red papule that increased in size over time. Patient also complained of episodic excessive sweating over the lesion. Examination revealed a hyperpigmented irregular plaque with a dusky-red hue measuring about 5 × 8 cm with geographical margins and smaller satellite lesions involving the left scapular region [Figure 1]. Histological examination of the lesion revealed multiple circumscribed lobules and aggregates of elongated and spindle-shaped cells throughout the dermis that demonstrated luminal differentiation at the center and at the margins [Figure 2]. The lumina were lined by large endothelial cells [Figure 3]. The overlying epidermis was mildly hyperplastic with irregular elongation of rete ridges and increased melanization of the basal layer.

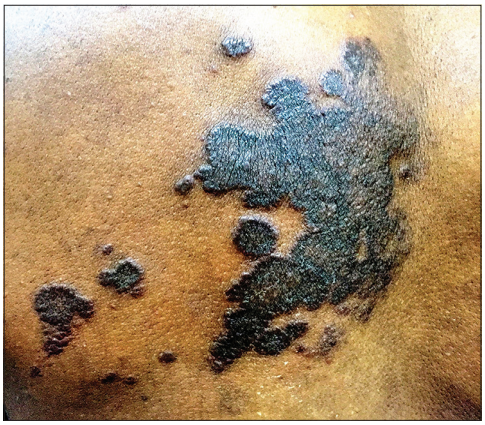

Figure 1: Irregular dusky red plaque with geographic margins and multiple satellite lesions on the left scapular region

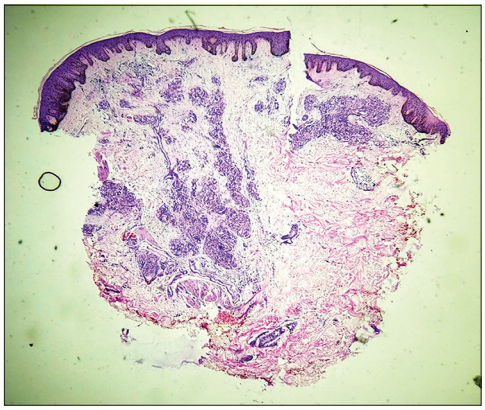

Figure 2: Multiple discrete lobules of cells with slit-like lumina in the center and at the margins are noted in the entire dermis (H and E, ×5)

Question

What is your diagnosis?

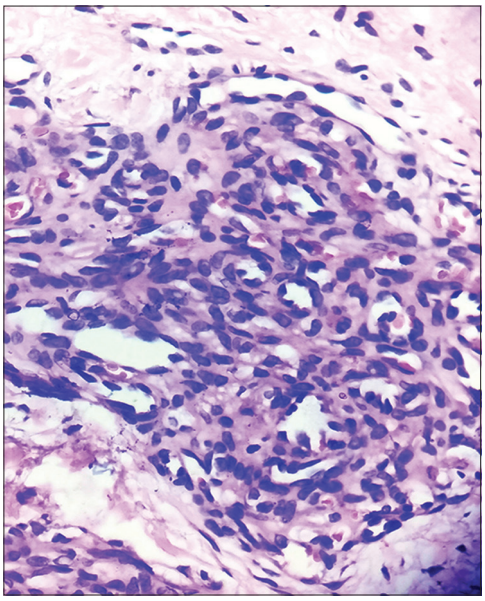

Figure 3: Higher magnification of an individual cellular lobule shows elongated and spindle-shaped cells with central and marginal luminal differentiation that are lined by large atypical endothelial cells (H and E, ×40)

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Quick Response Code:

Answer

Acquired (adult-onset) tufted angioma.

Discussion

Tufted angioma is an uncommon benign vascular tumor that derives its name from the characteristic histopathological feature of multiple tufts of aggregated elongated endothelial cells in the dermis.[1] Half of the lesions occur in the first year of life without any gender or racial predilection. Congenital tufted angiomas account for about 25% of the cases. The adult-onset forms are rare and have been reported sporadically. Less than 10% of the cases develop after 50 years of age, and the condition is very rare after 60 years.[2-4] Clinically, tufted angioma is characterized by a slow growing, dull red to violaceous or brown macules, plaques or nodules commonly involving the torso and proximal extremities. It can be associated with hyperhidrosis and/or lesional hypertrichosis. Although episodic lesional hyperhidrosis was present, hypertrichosis was absent in our case. Approximately 30% of the lesions have been reported to be tender.[5] Histopathologically, tufts of proliferating endothelial cells are scattered discretely throughout the dermis giving a "cannon ball" appearance. The cells are mostly elongated and spindle shaped with small slit-like vascular spaces mainly seen at the periphery that are lined by plump (atypical) endothelial cells protruding into the lumen. Immunohistochemically, the cells stain positively with endothelial markers such as CD31 and CD34 as well as lymphatic markers such as D2-40 and PROX1.[6]

The natural course of tufted angioma may be one with partial or complete resolution, persistence over many years, or development of Kasabach-Merritt syndrome.[5,7] In a series of thirteen cases, Osio *et al.,* identified three clinical patterns – tufted angioma without complications, tufted angioma complicated by Kasabach-Merritt syndrome, and tufted angioma without thrombocytopenia but with chronic coagulopathy. Hence, it is recommended that the initial work-up of patients should include a complete blood count and a more detailed evaluation for coagulopathy if platelet count is found to be less than $150 \times 10^3/\mu L$.[8] Kasabach-Merritt syndrome occurs in about 10% of the pediatric tufted angiomas, while its exact incidence in adults is unknown.[9] There are no definitive treatment guidelines for tufted angioma, and the choice of treatment modality is principally guided by the size, morphology, and location of the tumor, as well as presence or absence of complications like Kasabach-Merritt syndrome. In congenital and early onset cases, a careful periodic observational approach seems reasonable as spontaneous resolution occurs commonly in such instances. Several treatment modalities have been advocated when indicated and include surgical excision, pulsed dye lasers, and pharmacological measures such as systemic corticosteroids in tapering doses, vincristine, and interferon alfa.[10] In symptomatic patients where surgery is not feasible, aspirin 5 mg/kg/day is the first-line treatment.[11]

The differential diagnoses of tufted angioma include other vascular proliferative lesions such as congenital and infantile hemangiomas or vascular malformations in congenital and early-onset cases, and kaposiform hemangioendothelioma or Kaposi sarcoma in adult-onset cases.[12] Histologically, tufted angioma resembles glomeruloid hemangioma.[13]

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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