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Satellite lesions in tufted angioma: Atypical presentation

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Abstract
Tufted angioma (TA) is a rare vascular neoplasm. It can be congenital or acquired. Its clinical presentation is of a solitary angiomatous tumor. Multifocal lesions have been rarely reported in the literature. In addition, tufted angioma can be associated with Kasabach–Merritt phenomenon, though rare, causes complications and hence needs to be recognized. We report a case of multifocal tufted angiomas with a primary lesion and 2 satellite lesions for the rarity of the condition.

Keywords: tufted angioma, vascular, satellite lesion, multifocal

Introduction
Tufted angioma (TA) is a rare vascular neoplasm [1]. The term tufted angioma is due to the dense agglomerates of endothelial cell lobules and capillaries on histology. It can be congenital or acquired. Its clinical presentation is of a solitary angiomatous tumor, usually located on the neck, upper trunk and extremities [2]. Multifocal lesions have been rarely reported in the literature. These tumors follow a benign course of angiomatous proliferation, without any evidence of malignancy [1]. In addition, tufted angioma can be associated with Kasabach–Merritt phenomenon, though rare, causes complications and hence needs to be recognized [3].
We report a case of multifocal tufted angiomas with a primary lesion and 2 satellite lesions for the rarity of the condition.

Case Report
A 6-year-old boy has consulted for a reddish painful papule of the left shoulder, which had first noticed 6 mounths before the consultation.The lesion gradually became bigger, elevated and painful. One month later, new two lesions appeared near to the first one. The lesions have remained stable for the preceding several months. There was no history of bleeding from the lesions or any other body site. Family history and past medical history was unremarkable.
On examination, three erythematous papules were seen over the left shoulder. The first lesion was about 3 cm diameter, well-marginated with a pale center. Two satellite lesions of about 8 mm were seen around the first lesion (figure 1). There was neither hypertrichosis nor increased sweating. Dermoscopy examination showed numerous tiny red peripheric lacunae (figure 2).
No other systemic or cutaneous abnormalities were detected. Routine laboratory tests, blood coagulation profile, and serum biochemistry panel were normal.
Histopathological examination revealed mesenchymal tumor proliferation of a vascular nature involving the entire dermis. This vascular proliferation is made up of small, well-defined lobules arranged in the dermis, giving the appearance of small tufts. The sweat glands are slightly hyperplastic (figure 3). Based on the clinical findings and characteristic histology, a diagnosis of tufted angioma was made. After discussion about the possible modalities of treatment and the chance of spontaneous regression, the parents preferred regular follow-up without any active intervention and the child was kept under observation.

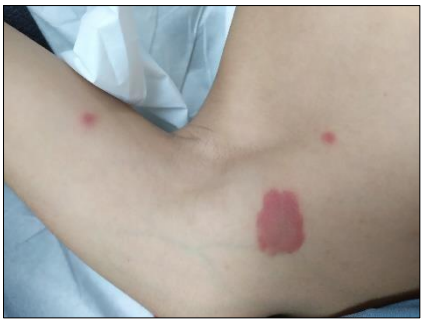

Fig 1: 3 Red papules of the left shoulder

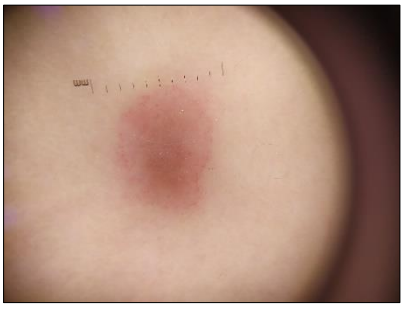

Fig 2: Dermospic image with tiny red peripheric lacunae

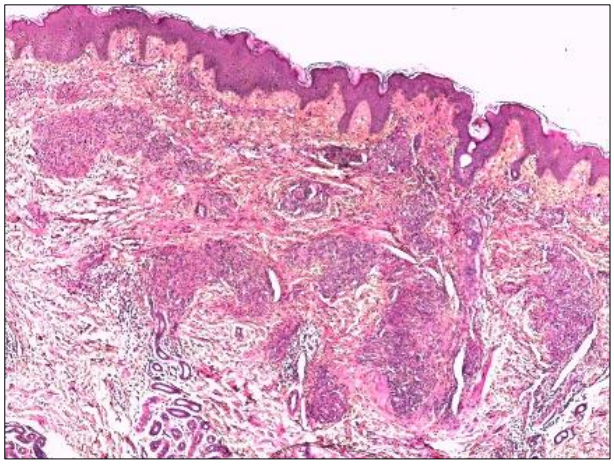

Fig 3: Histologic image with vascular proliferation and well-defined lobules arranged in the dermis.

Discussion

TA was described for the first time by Wilson Jones in 1976 as an acquired vascular proliferation of peculiar histological characteristics and reports of additional cases had helped better characterization of this clinicopathologic entity [1]. The same condition was named as angioblastoma by Nakagawa in 1949 in Japanese literature [3]. Most cases of angioblastoma reported have been from Japan [4].

Congenital tufted angiomas account for about 25% of the cases [5]. It commonly presents in infancy, or early childhood [1]. However, adult-onset cases have been reported also with an onset at the age of 53 [4].

TA usually presents as red a macule, papule or plaque over the upper trunk, neck and proximal part of the limbs; involvement of other locations like face, oral mucosa and lip is also known [1]. It can be associated with hyperhidrosis and/or lesional hypertrichosis wich is absent in our case [5]. Only a few reports have been published showing the dermoscopic features of TA. Ma *et al*.4 described a case without red lacunae shown on dermoscopy [6]. In another case report, dermoscopy revealed numerous tiny red lacunae separated by thin, whitish linear septa. TA should be differentiated from Kaposiform hemangioendothelioma, juvenile capillary, or strawberry angioma, haemangiopericytoma, glomeruloid haemangioma, or pyogenic granuloma [1].

Tufted angioma is sometimes misdiagnosed, thus histopathologic examination and sometimes immunohistochemistry are required for confirmation [3]. It has a distinctive histopathological pattern characterized by the vascular tufts of densely packed capillaries, randomly scattered throughout the dermis in a so called "cannonball" configuration. There are crescentic spaces surrounding the vascular tufts and lymphatic-like spaces within the tumor stroma. The epidermis is normal in most cases. Immunohistochemical stains show strong positivity for Ulex uropaeus I Lectin and EN4 and unlike infantile hemangioma, negative staining for GLUT [1].

Kasabach-Merritt syndrome occurs in about 10% of the pediatric TA, while its exact incidence in adults is unknown [5]. It is Essential to obtain a complete blood cell count and coagulation function test when suspecting a child with tufted angioma complicated with Kasabach–Merritt phenomenon. Owing to the high mortality rate of this condition, it should be intervened as early as possible [3].

In a series of thirteen cases, Osio *and 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 [5]. TA usually pursue a persistent course, but spontaneous regression may occur in a proportion of cases [1].

Currently, there are no treatment guidelines, and the therapy for tufted angioma is limited [3]. 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 [5].

Several treatment modalities have been advocated when indicated and include surgical excision, pulsed dye lasers [7]. Pharmacological measures such as systemic corticosteroids in tapering doses, vincristine, and interferon alfa are also proposed. In symptomatic patients where surgery is not feasible, aspirin 5 mg/kg/day is the first-line treatment [5].

Recently topical tacrolimus and timolol topic have shown good resultats in case reports [8, 9]. Some authors believe the lesion should only be monitored due to the possibility of spontaneous regression of these cases [2].

Kasabach–Merritt phenomenon needs aggressive treatment, such as corticosteroids, vincristine or interferon-α [3].

Conclusion

In conclusion, we have reported a rare case of tufted angioma with satellite lesions in a child as an atypical clinical presentation. Therefore the possibility of multiples lesions should be kept in mind.

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