A tufted angioma

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ABSTRACT

Tufted angioma (TA) is a rare, benign, cutaneous angiomatous proliferation. It is more common in children, usually presenting as red-purple painful plaques on the trunk. We describe here a TA observed at nine months of age, appearing initially over the retroauricular area, gradually extending to involve skin of neck and trunk by two years of age. This case of a large TA (7 × 12 cm) in an Indian male child is reported here due to its rare presentation.

Key words: Angiosarcoma, Kasabach–Merritt syndrome, tufted angioma

INTRODUCTION

Tufted angioma (TA) is a rare vascular tumor most commonly localized to the skin and subcutaneous tissue, characterized by slow angiomatous proliferation. The term TA was coined because of the characteristic dense clumps and lobules of endothelial cells and capillaries observed on histology.[1] It is a variant of capillary hemangioma and supported by the finding of characteristic crystalline lamellae in both of them.[2] It may be evident at birth, but usually develops during late infancy or adolescence. Acquired TA has been reported in immunocompromised adults.[3]

CASE REPORT

A 2-year-old boy presented with coalescing, red-purple, firm plaques with irregular borders and few superimposed papules extending from the right retroauricular region to his neck and upper trunk.

His mother gave a history that the child initially had a small erythematous macule behind the right ear at 9 months of his age, which gradually progressed to multiple plaques, extending on to the neck and front of the chest. On examination, the plaques were deep red in color with few areas of violaceous discoloration and telangiectasia at the margins involving approximately 7 × 12 cm area from right ear to right anterior chest [Figure 1 and 2]. The lesions on palpation were tender with mild induration. Routine investigations including hemogram, liver and renal function tests, and chest X-ray were within normal limits. Histopathology revealed a normal epidermis with the dermis showing proliferating capillaries forming into nodules lined by endothelial cells [Figure 3 and 4]. A few vascular lumina showed red blood cells. A sparse lymphohistiocytic infiltrate was noted in the upper dermis. There was no evidence of mitoses. The features were consistent with a diagnosis of TA.

DISCUSSION

TA, also called angioplasty of Nagakawa, is a very rare pattern of angioma. Approximately 25% are congenital and 50% appear in the first year of life.[4] A few cases occurring on the oral mucosa and in adults have been reported. Sex distribution is equal without any racial predilection. Some cases have been reported in a familial pattern and a few in pregnancy which resolve after delivery. It usually starts as a small macule, which resembles a port wine stain and progresses to deep red to purple plaques or nodules of 2–10 cm size.[5] In the present patient, the TA was larger, involving 7 × 12 cm area. The most common sites of involvement are the neck, shoulder, or upper trunk, and occasionally the proximal limbs. On histopathology, TA shows multiple scattered lobules or tufts having a cannonball appearance. TA is a benign condition, pain and tenderness are common associated symptoms, and hyperhidrosis is a frequent finding that occurs in 30% of patients.[6] In a review of 13 cases of TA, the author observed three different clinical patterns: TA without

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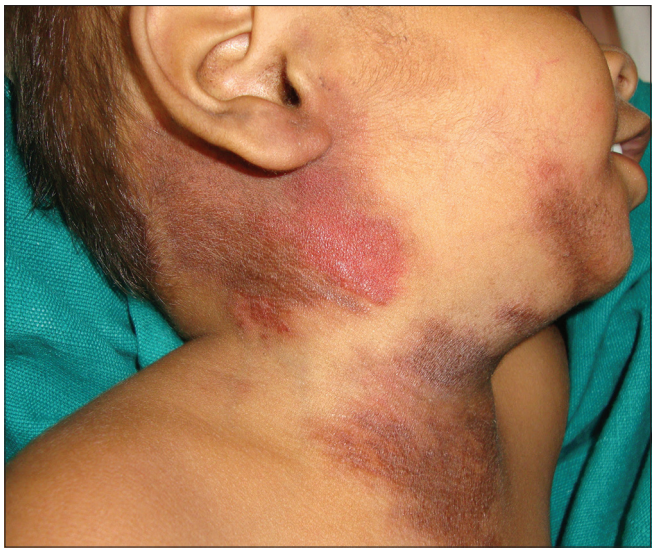

Figure 1: Red to purple colored plaque of tufted angioma over the right side of neck

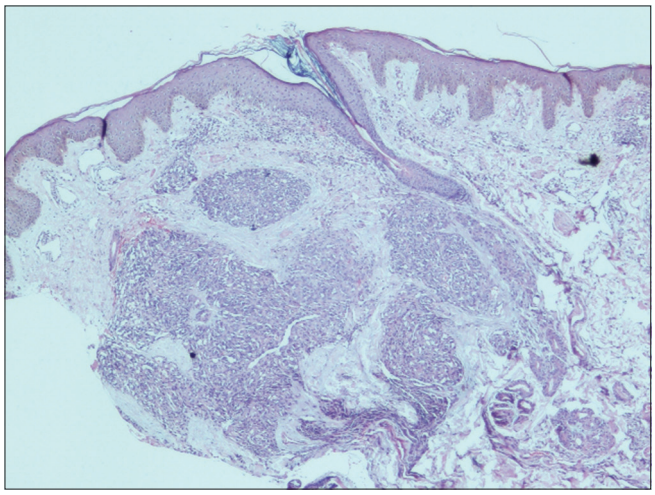

Figure 3: Photomicrograph showing lobules of proliferating capillaries (H and E, 10 × 10)

complications, TA without thrombocytopenia but with chronic coagulopathy, and TA complicated by Kasabach-Merritt syndrome with thrombocytopenia. Of these first pattern is most common.[7] Spontaneous regression commonly occurs when onset is before six months of age.[8] When the onset is later, partial spontaneous regression of TA may occur but complete disappearance is extremely rare. Super potent topical corticosteroid is the mainstay of treatment. Pulsed dye laser has been reported as an option for treatment of TA associated with pain. The other modality is complete surgical excision, useful only for smaller lesions. Our patient was given moderately potent topical corticosteroid and the lesion regressed partially.

CONCLUSION

Our patient had classic clinical features of indurated red to purple plaques and typical histological picture of nodules of

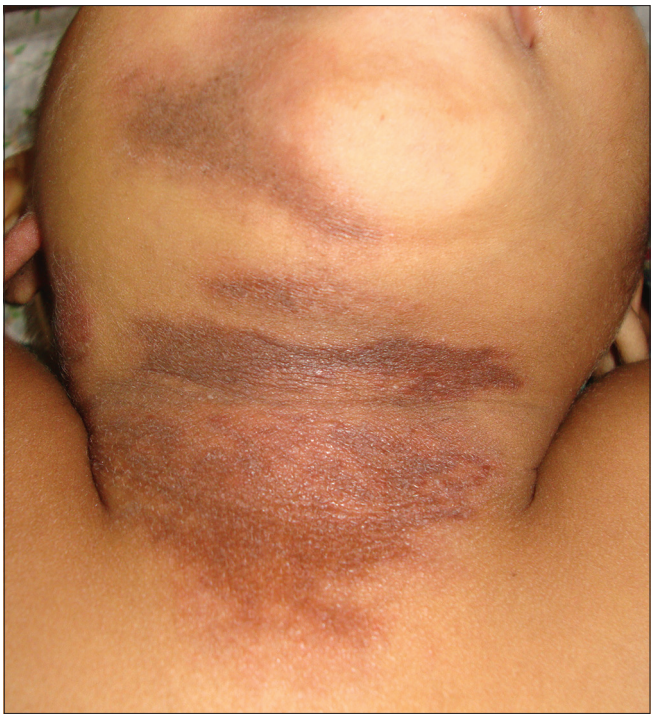

Figure 2: Tufted angioma involving neck and extending on to chest

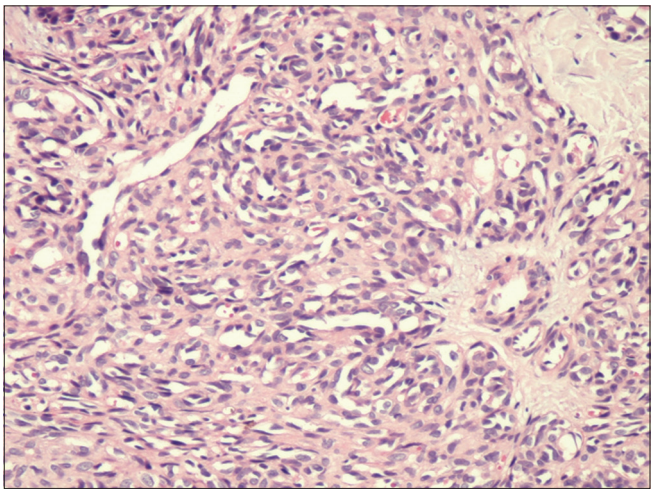

Figure 4: Higher magnification of the proliferating capillaries (H and E, 10 × 40)

proliferation of capillaries lined by endothelial cells and no mitoses, consistent with a diagnosis of TA. This case of TA involving large areas of skin over the head, neck, and chest is reported for its rarity.

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