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Case Report

Acquired giant tufted angioma: a rare entity

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Abstract Tufted angioma is an uncommon benign vascular tumour encountered usually in infancy and early childhood. Lesions are mostly confined to the trunk and face and generally not larger than 10 cm in diameter. Our case was about an 11-year-old boy who presented with a very large, enhancing, reddish brown coloured, tender plaque on loin. Histopathology revealed circumscribed, multiple foci of closely set vascular channels giving the canon ball appearance suggestive of tufted angioma. Giant tufted angiomas are rare and hence reported.

Key words
Tufted angioma.

Introduction

Tufted angioma (TA) is a rare benign cutaneous vascular neoplasm. These tumours are acquired mostly during prepubertal age and pursue a benign course of slow angiomatous proliferation without any aggressive behavior or metastasis.[1] TA typically presents as dull red or red brown patches and plaques of 2-5cm over neck, upper trunk and proximal aspect of limbs. Tumours exceeding 10 cm is a rarity. These are known as giant tufted angiomas. Our case was an 11-year-old boy presenting with a large indurated plaque over right lumbar region which was diagnosed as tufted angioma on histopathology.

Case report

An 11-year-old child presented with a well-circumscribed, hyperpigmented, indurated plaque with an erythematous hue over right loin for 6 months (**Figure 1**). It started as a small area of thickened skin which slowly enlarged and over the last one month there had been a rapid increase in size of the lesion. It was found to be 15cm×11cm in size. The overlying surface was irregular and studded with numerous small papules. The skin was warm and indurated and could not be pinched. The lesion was non tender, non-compressible and no pulsation, thrill or bruit could be felt. The patient denied any history of trauma, bleeding or increased sweating from the plaque. No history of bleeding from nose or gum could be recounted by the child. Significant family history was not present. Skin at other sites, hair, nail or mucous membrane was unremarkable. Systemic examination was non contributory. Connective tissue hamartoma, morphea and panniculitis were considered as clinical differential diagnosis. Complete heamogram showed normal finding except mildly reduced platelet count. Prothrombin time APTT, BT, CT and other routine biochemical tests were within normal limits. Chest X ray and USG of abdomen was normal. Colour Doppler study of right lumbar region showed a vascular malformation with dilated capillaries in dermis, subcutis and extending up to intercostal muscles (**Figure 2**).

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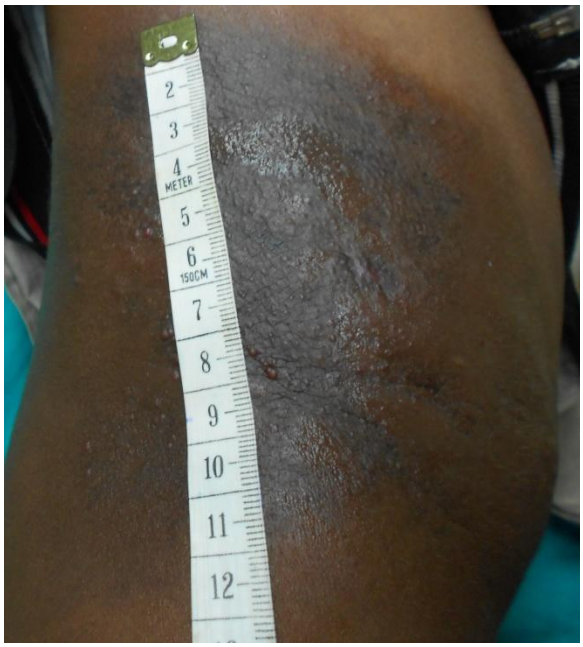

Figure 1 Giant tufted angioma on right loin of the patient.

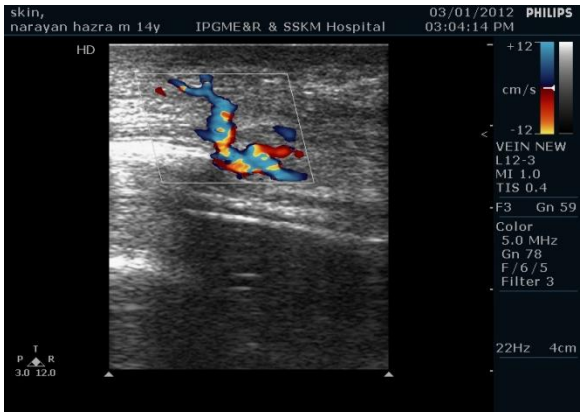

Figure 2 Colour Doppler showing invasion of the muscle planes by the vascular lesion.

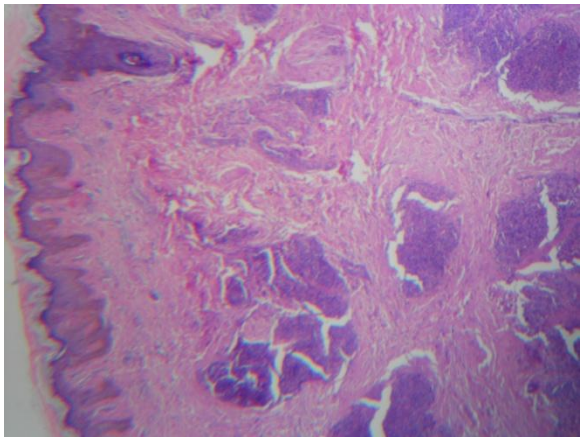

Figure 3 Histopathology showing tufts of capillaries in typical 'cannonball' distribution.

Histopathological examination of the skin biopsy showed multiple cellular lobules in dermis composed of proliferating capillaries and endothelial cells arranged in typical 'cannon ball' distribution (**Figure 3**). A diagnosis of tufted angioma was made and the patient was referred to the Surgery department for necessary management.

Discussion

Tufted angioma has been described in the literature under different names including Nakagawa's angioma, Nakagawa's angioblastoma, progressive capillary hemangioma, and acquired tufted angioma of the skin and subcutaneous tissue.[2] It is a rare, benign vascular tumour occurring most commonly in prepubertal children.[3] A few cases occurring at unusual sites like oral mucosa in adults have been reported.[4] The lesion of TA enlarges slowly over 5 months to 10 years after which no further growth occurs. The skin lesions occur without a history of preceding trauma, except for one case where tufted angioma had followed an arthropod bite.[5] Clinically, TA appears as a dusky red or purple patch or plaque with superimposed angiomatous papules that predominantly occurs on upper thorax, neck and shoulder. They can reach upto 20cm in diameter and may cause diagnostic confusion with port-wine stain, connective tissue naevi, smooth muscle hamartoma. Hyperhidrosis on the surface may be associated and when it occurs it corresponds histologically with an area of abundant collagen.[6,7] Occasionally TA may be associated with Kasabach-Meritt syndrome which causes platelet trapping and consumptive coagulopathy.[1] Magnetic resonance imaging (MRI) studies are ideal for evaluating the depth of invasion of TA. In our case, as the patient could not afford MRI, colour Doppler study, a cheaper alternative was performed which

indicated intercostal muscle invasion by the tumor.

TAs have a specific and distinctive histopathological pattern. There are scattered lobules or tufts composed of hypertrophied endothelial cells, pericytes and capillary vessels in reticular dermis, giving the characteristic cannon ball appearance. Occasionally the tufts coalesce to form irregular vascular tracts that invade into deep fascia and muscle.[8] Mitosis and cellular atypia are absent. Immunohistochemistry shows strong positivity for Ubx European I lectin and EN_4 and unlike infantile haemangioma, negative staining for GLUT-1.[7] TA and Kaposiform hemangioendothelioma (KH) sometimes show overlapping histopathological features. Immunohistochemistry is helpful for distinguishing the two conditions.

Different modalities of treatment have been tried for TA. The recurrence rate is very high after surgical excision. Soft X-ray, pulse dye laser, IFN-α and high dose corticosteroid are other options and they have shown variable response. A case of giant TA successfully treated with radiation has also been reported.[9]

Tufted angioma is a rare entity and the giant variety is even rarer. Thus, the case is reported here to emphasize the importance of considering TA in the differential diagnosis of any acquired vascular lesion.

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