Acquired tufted angioblastoma in an immunocompetent patient


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Abstract

Acquired tufted angioma (ATA) is a rare benign vascular tumor, most commonly localized to skin and subcutaneous tissues. Its name is based on its specific histologic pattern, characterized by lobular arrangement of densely cellular capillaries, grouped in glomerular pattern looking like ‘cannon-ball’. These vascular tufts are comprised of endothelial cells and associated pericytes. Angioblastoma are often associated with immunocompromised state. We report a case of acquired painful solitary angioblastoma in immunocompetent adult individual.

Key words

Acquired, angioblastoma, vascular tumor, immunocompetent.

Introduction

Tufted angioblastoma is rare vascular neoplasm localized in skin and vascular tissue occurring primarily on trunk and extremity of the children.¹ This angioma is ill-defined dull red macule or plaque with a mottled appearance. This tumor pursues a benign course without any evidence of metastasis.² There is no sex or gender predilection for the disease. The lesions are usually asymptomatic but rarely paroxysmal painful episodes were reported.³ Angioblastoma is often associated with immunocompromised state. We report a case of acquired painful solitary angioblastoma in an immunocompetent adult individual.

Case report

A 40-year-old man presented to Dermatology OPD of School of Tropical Medicine with a solitary elevated painful scaly skin lesion in front of right ear for 6 months. There was no history of trauma or bleeding, increased sweating or discharge from the lesion. There was no family history of similar illness.

Examination revealed an erythematous scaly plaque with irregular border, measuring 5 cm in the greatest dimension, in front of right ear (Figure 1). The lesion was firm in consistency and was nontender. There was slight rise of local temperature. Examination of nail, hair, mucosa and other part of skin was normal. Systemic examination was also not contributory.

His hemogram, blood biochemistry, bleeding time, clotting time and prothrombin time were normal. HIV ELISA was nonreactive and CD4 count was 720 cell/mm³. His chest X-ray and ultrasonography of whole abdomen were normal. Punch biopsy from the lesion showed single lobule consisting of multiple capillaries with bloodless lumen which is surrounded by crescent shaped vascular channel giving ‘cannonball’ appearance.
Based on the clinical and histopathological feature we diagnosed the case as angioblastoma.

**Discussion**

Tufted angioma usually develops in infancy and early childhood. Usually it is solitary but multiple lesions are reported. Clinically, angioblastoma present as reddish macule or plaque with cluster of superimposed small angiomatous papules; annular configuration with central depression may also occur. The common site of occurrence are head, neck and upper trunk. The lesions are firm to hard in consistency and rarely tender. The lesions are usually sporadic although familial cases have been reported. Angioblastoma can occur in eruptive form. Chup *et al.* described an acquired tufted angioma following liver transplantation. It may be associated with Crohn’s disease and pregnancy and HIV patients. The differential diagnosis of angioblastoma are pyogenic granuloma, kaposiform hemangioendothelioma (KHE), hemangiopericytoma, strawberry angioma etc. Hypertrichosis has been noted with angioblastoma. Kasabach-Merritt syndrome (KMS) may be associated with angioblastoma. Confirmation of diagnosis of angioblastoma is by histopathological examination which shows vascular tufts of densely packed capillaries surrounded by crescent shaped blood vessel which is known as ‘cannonball’. Magnetic resonance imaging (MRI) is helpful in the evaluation of depth of invasion of tufted angioma. On suspecting KMS complete hemogram and thorough coagulation profile (bleeding time, clotting time, prothrombin time, PTT, APTT), fibrin degradation product and D-
dimers should be evaluated. Tumours with features of tufted angioblastoma and kaposiform hemangioendothelioma have been described. Transformation between these two tumours has also been recorded. Immunostaining is helpful in distinguishing between these two tumours. Angioblastoma shows proliferation of CD34 positive endothelial cells with few actin positive cells whereas KHE shows CD34 staining cell only in luminal endothelial cells. In most of the cases angioblastoma is slowly progressive and complete regression is extremely rare. The treatment modalities of angioblastoma include potent topical steroids, complete surgical excision, cryotherapy and radiotherapy. High dose interferon alfa-2 has also been tried with variable result. Pulse dye laser may be an effective treatment modality for angioblastoma.

This case is reported because of late age of presentation, symptoms during presentation and to emphasize that angioblastoma should be considered in the differential diagnosis of acquired vascular tumours.

References