

Acquired tufted angioma - a case report and review of the literature

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Acquired tufted angioma is an unusual vascular proliferation first described by Wilson Jones in 1976 and further characterised in 1989^{1,2}. Lesions previously described as progressive capillary haemangioma³ and in the Japanese literature as 'angioblastoma' (Nakagawa)⁴ appear to be identical to the acquired tufted angioma.

This lesion presents clinically as a slowly spreading erythematous macules and papules, predominantly in the neck and upper trunk. There may also be raised papules resembling a pyogenic granuloma. With the association of Kaposi's sarcoma (KS) in patients with acquired immunodeficiency syndrome (AIDS), pathologists are justifiably suspicious when diagnosing acquired vascular lesions specially in the adolescents and young adults. This underlies the importance of defining new vascular lesions, specially the rarer entities, in this group where KS may be wrongly considered. We report a case of acquired tufted angioma in a 42 year old male.

Case history

A 42 year old male presented with multiple spreading 'warty' papules in the front and right side of the neck region over a six month period. The lesions were non tender. He did not give a history of fixed vascular blemishes since infancy. There was no family history of similar lesions.

One lesion was excised under local anesthesia as an outpatient procedure and sent for histological assessment in formol saline.

Histology

The low power examination showed a focal arrangement of multiple separated cellular lobules within the dermis (Fig 1). The capillary sized vessels occurred in discrete ovoid tufts in a background of normal dermal collagen. Some lobules bulge the walls of dilated thin walled vascular channels (Fig 2, arrow) which are within the lobules or at the periphery. Each lobule is composed of aggregates of endothelial cells that are concentrically whorled along a preexisting vascular plexus (Fig 2). Small capillary lumina were seen within the lobules. There was no cellular atypia, papillary processes or mitoses. There was no inflammation, edema or hemosiderin.

This lesion lacked the epidermal colarette formed by elongated rete ridges which is seen in pyogenic granuloma.



Figure 1. Focal arrangement of multiple separated cellular lobules within the dermis (haematoxylin and eosin x 600)

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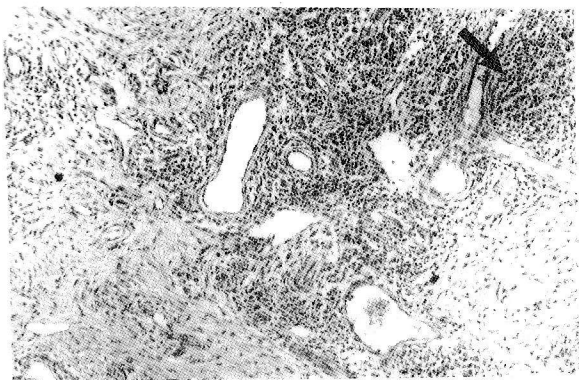


Figure 2. Lobule composed of aggregates of endothelial cells concentrically whorled along a preexisting vascular plexus with small capillary lumina. Some lobules bulge the walls of vascular channels (arrow) (haematoxylin and eosin $\times 600$)

Discussion

The histological features with the 'cannon ball' pattern is characteristic of an acquired tufted angioma⁵. The slow clinical course and the histological features indicate a benign course², although it has been confused with a low-grade angiosarcoma in the past¹. Our patient was thought to have a 'warty' lesion clinically, and not a vascular abnormality. There have been reports of clinical queries of connective tissue abnormalities or granulomas² and our case adds to this category of 'non-vascular' queries on clinical examination. Our patient did not show the red, superficial component usually associated with angiomatous lesions and though it may be due to our skin colour, this feature has also been described in white skin². The relative bloodlessness of the vascular tufts and the predilection for lower dermal involvement may explain the lack of a 'vascular blush'.

This condition usually stops growing after some years but there is usually no tendency to involute and no metastases have been reported^{3,6,7}. There has been no explanation as to why some histologically benign lesions should spread after remaining stable for some time².

Spontaneous regression in acquired tufted angioma is rare⁷. Soft radiation treatment has been recommended⁷. Satisfactory results have been reported with surgery, pulse dye laser, high dose systemic steroids and interferon alfa⁷.

Histologically this lesion shows some features of a strawberry angioma on one hand and pyogenic granuloma on the other⁶. However, the cannon ball like arrangement is not seen in either.

This entity may also need to be distinguished from endovascular papillary angioendothelioma of childhood and Masson's vegetant intravascular haemangioendothelioma. Both these conditions have papillary processes in addition to the cellular lobules. Endovascular papillary angioendothelioma of childhood contains papillae lined by atypical endothelial cells which protrude into vascular lumina. In Masson's vegetant intravascular haemangioendothelioma the papillary processes are composed of hyperplastic endothelium supported by fibrous stalks, and this is confined within vascular lumina.

The pattern of cellular nodules with peripheral dilated channels may resemble Kaposi's sarcoma. However, the lobules lack the characteristic interlacing fascicles of spindle cells lining slit like vessels seen in an Kaposi's sarcoma. Kaposi's sarcoma usually has a plasma cell infiltrate, which is not seen in an acquired tufted angioma.

Immunohistochemistry has demonstrated weak positivity or negativity for F-VIII related antigen but positivity with smooth muscle actin in the capillary tufts indicating pericytes. Strong positivity with Ulex europaeus I lectin and EN4 indicate the endothelial nature⁷. Ultrastructural studies have demonstrated characteristic crystalloid inclusions within the endothelial cells⁴ in addition to Weibel Palade bodies⁷.

We conclude that acquired tufted angioma is a distinctive, rarely recognised condition² that is of importance in the differential diagnosis from other acquired vascular lesions in young persons. Despite the progressive spread

this angioma behaves in a benign manner. Histology is essential for the diagnosis and to exclude the more sinister conditions.

References

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