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 haemangioma[3] and in the Japanese literature as 'angioblastoma (Nakagawa)[4] apperas 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 entites, in this group where KS may de 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 haemosiderin.

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

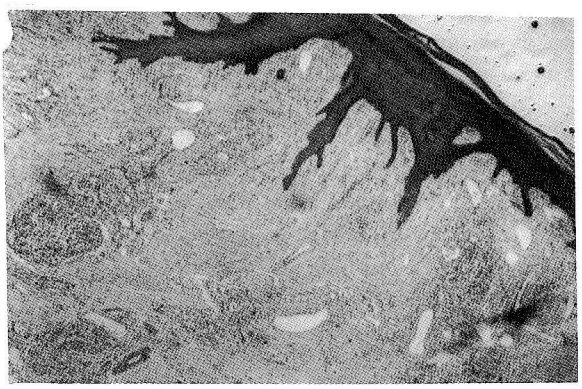

Figure 1. Focal arrangement of multiple separated cellular lobules within the dermis (haematoxylin and eosin x 600)

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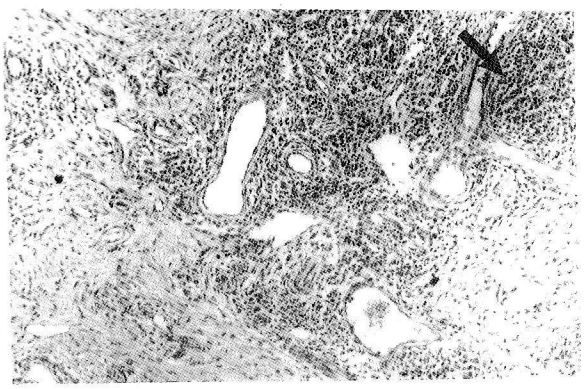

Figure 2. Lobule composed of aggregates of endothelial cells concentrically whorled along a preexisting vascular plexus with small capillary lumina. Some lobules bulge the walls of vascular channels (arrow) (haematoxylin and eosin x 600)

Discussion

The histological features with the 'cannon ball' pattern is characteristic of an acquired tufted angioma[5]. The slow clinical course and the histological features indicate a benign course[2], although it has been confused with a low-grade angiosarcoma in the past[1]. 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 granulomas[2] 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 skin[2]. 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 time[2].

Spontaneous regression in acquired tufted angioma is rare[7]. Soft radiation treatment has been recommended[7]. Satisfactory results have been reported with surgery, pulse dye laser, high dose systemic steroids and interferon alfa[7].

Histologically this lesion shows some features of a strawberry angioma on one hand and pyogenic granuloma on the other[6]. However, the cannon ball like arrangement is not seen in either.

This entity may also need to be distinguished from endovasular 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 edothelial nature[7]. Ultrastructural studies have demonstarted characteristic crystalloid inclusions within the endothelial cells[4] in addition to Weibel Palade bodies[7].

We conclude that acquired tufted angioma is a distinctive, rarely recognised condition[2] 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 bengin manner. Histology is essential for the diagnosis and to exclude the more sinister conditions.

References

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