Recurrent aneurysmal fibrous histiocytoma

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Aneurysmal fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma that results from blood vessel proliferation and haemorrhage into a fibrous histiocytoma. The resulting lesion has a very different clinical appearance, hence the potential confusion with other skin lesions. This report describes the case of a 48 year old woman with a recurrent fibrous histiocytoma with prominent vasculature, which over a three year period recurred on two occasions, showing more progressive features of the aneurysmal variant. In addition, squamous lined cysts were present within this tumour, a finding that has not been described previously. The histological features of this rare lesion and the importance of the differential diagnosis from other similar appearing malignant lesions will be discussed.

Cutaneous fibrous histiocytoma is a benign dermal tumour composed of fibrohistiocytic cells. The principal variants are fibroblastic and histiocytic, although numerous morphological variants occur, including clear cell, atrophic, epithelioid, cellular, and finally aneurysmal fibrous histiocytoma (AFH). This is an uncommon lesion, first described in 1981,¹ and few cases have been reported.² Hence, we highlight the importance of separating this entity from similar malignant skin lesions.

CASE REPORT

A 48 year old woman presented with a long standing skin lesion on the calf, which had been progressively increasing in size and was painful. After excision, histology showed a spindle cell and histiocytic lesion with a storiform pattern

Figure 1 Fibrohistiocytic cellular proliferation containing numerous blood filled cysts, the largest measuring 8 mm. Stromal haemosiderin deposition was confirmed by Perls staining. Haematoxylin and eosin stain; original magnification, $\times 100$.

Take home messages

- We describe a case of aneurysmal fibrous histiocytoma (AFH), which was aggressive and recurrent
- It is important to distinguish this benign lesion from similar appearing malignant skin lesions
- Squamous lined cysts were present within this tumour, a finding that has not been described previously

containing foamy histiocytes admixed with large numbers of dilated blood vessels. The lesion was reported as a cellular dermatofibroma with prominent vasculature.

However, the lesion recurred twice at the same site and the final excision was performed down to the fascia. The lesion measured 2.5 cm and contained a well defined cystic tumour. Microscopic examination showed a resemblance to the original lesion, although it now contained cystic, blood filled spaces, the larger of which were lined by squamous epithelium. The smaller cysts had no lining, but were bordered by intervening spindle shaped cells extending to the subcutaneous tissue (figs 1 and 2).

Immunohistochemistry for CD34 confirmed the absence of an endothelial lining around the small blood filled spaces and cytokeratin staining confirmed the presence of squamous epithelium around the larger cysts (figs 3 and 4).

The features were consistent with the rare aneurysmal variant of fibrous histiocytoma. The patient required skin grafting one month after the third excision.

Figure 2 The cysts lacked an endothelial lining and were bordered by epithelioid and spindle shaped cells with eosinophilic cytoplasm showing a storiform pattern. Pleomorphism was mild and occasional mitoses were present. Haematoxylin and eosin stain; original magnification, $\times 400$.

Figure 3 The larger cysts were bordered by a squamous lining beneath which were fibrohistiocytic cells. Haematoxylin and eosin stain; original magnification, $\times 200$.

DISCUSSION

AFH is characterised by fibrohistiocytic cellular proliferations containing blood filled spaces lacking endothelium.¹ Although the pathogenesis is unknown, it may be caused by extravasation of erythrocytes from capillaries leading to the cystic blood spaces.³ However, the presence of the larger squamous lined cysts has not been described previously and could be misleading.

Clinically, this may be confused with malignant melanoma or haemangioma. Histologically, there is frequent confusion with other vascular tumours.² Cutaneous angiosarcoma usually occurs on the face or scalp of elderly men and contains atypical endothelial cells that separate collagen bundles.⁴ Kaposi's sarcoma may be difficult to distinguish from AFH, but is immunoreactive for CD34 and lacks fibrohistiocytic cells.⁵ "Angiomatoid" malignant fibrous histiocytoma is similar but occurs in deeper subcutaneous tissue, muscle, or periosteum.^{6,7}

"The presence of the larger squamous lined cysts has not been described previously and could be misleading"

In conclusion, we describe an aggressive recurrent AFH with squamous lined cysts and highlight the importance of distinguishing this benign lesion from similar appearing malignant skin tumours. Finally, an unusual histological change is described for this uncommon but distinctive clinicopathological entity.

Figure 4 The squamous lining lacked a granular layer and its epithelial nature was confirmed by cytokeratin (AE1/AE3) immunoreactivity. Haematoxylin and eosin stain; original magnification, $\times 400$.

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