Although 14% of our h-caldesmon positive AFX recurred, a much higher value than the usual 1% recurrence rate,[1] it is insufficient to consider if similar to that described in UPS, h-caldesmon immunoreactivity in AFX could indicate a higher potential for recurrence.

In summary, AFX myofibroblastic tumour cells can upregulate h-caldesmon expression under certain circumstances such as tissue remodelling secondary to epidermal ulceration. Therefore, a panel of muscle markers is needed to confidently differentiate AFX and leiomyosarcoma.

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Localised lymphoedema forming a papillated lesion on the scalp

Sir,

The lymphatic system is an important network of vasculature which removes waste products, transports chyle and is a critical component of the immune system. Mechanical failure of the lymphatic system results in increased protein-, lipid- and debris-rich interstitial fluid and underlies the development of lymphoedema.[1] In this setting a fibroinflammatory process ensues which initially presents as soft, swollen tissue but then progresses to thick and stiffened skin, with associated surface changes such as hyperkeratosis, acanthosis and papillomatosis. The stereotypical presentation of this phenomenon involves entire limbs or the genitalia ('elephantiasis') and is well described, particularly in the setting of filariasis in the developing world and post-lymphadenectomy or other treatment for malignancies in Western countries.[2] This condition can be the cause of significant morbidity for affected patients. Less well recognised is the same process involving smaller, more localised regions of the skin ('localised lymphoedema'), which can present with polyps, plaques, pendulous swellings or sarcoma-like masses.[3] The recognition of these lesions is critical to avoid over-diagnosis of more significant lesions, as well as to enhance our appreciation of the potential aetiological role these more restricted areas of lymphoedema may play in a number of other cutaneous lesions. We have recently encountered an unusual case of this process involving the scalp, which we report herein.

A 28-year-old male presented to his dermatologist with a prominent papillomatous plaque on the posterior scalp (Fig. 1). The lesion measured approximately 40 × 20 mm, and was characterised by a lobulated surface with deep fissures between the individual lobules. The dermatologist noted that the lesion was continuous with a cutaneous vascular malformation which extended down the posterior aspect of the neck. The patient confirmed the congenital nature of this malformation. However, he had only noticed the papillomatous lesion some 3 weeks prior to presentation, in the context of a magpie strike to the area. He was otherwise well with no neoplastic history and no significant family history.

An initial shave biopsy was performed for diagnostic purposes, which we favoured to represent an 'angiofibroma'. Subsequently a more extensive shave excision was performed, primarily for cosmesis. Histological examination of this specimen revealed multiple fragments of skin adopting

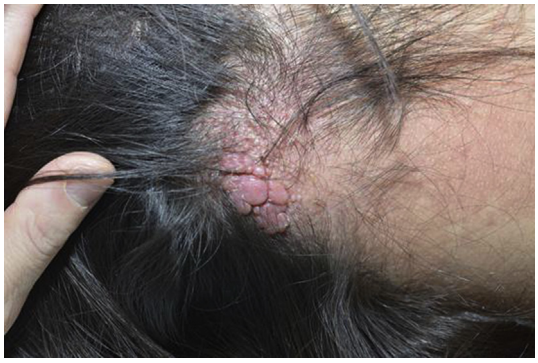

Fig. 1 The clinical appearances of the lesion on the scalp. It presented as a papillomatous, skin-coloured plaque.

a verrucous, polypoid architecture (Fig. 2A). There were foci of mild epidermal acanthosis, including scattered areas resembling dilated follicular infundibula with associated suppurative inflammation. The dermis was fibrotic, with evenly spaced, cytologically bland spindled cells. In addition there were increased vascular structures, including vessels showing irregular dilatation (Fig. 2B). These vessels were lined by a single layer of endothelium, some with a few layers of perivascular cells. While some were undoubtedly blood vessels, immunohistochemical staining for D2-40 confirmed that many represented dilated lymphatics (Fig. 2D). A mild, focal inflammatory infiltrate which included plasma cells was also present (Fig. 2C). This was predominantly superficial and perivascular in distribution. Dermal inflammation was also seen in association with the area of surface suppuration. The constellation of histological features was consistent with an area of localised lymphoedema. In addition, there was a focus of more conventional dermal scarring. While this was superficially similar to the rest of the lesion, a subtle distinction could be discerned in the form of more horizontally arrayed collagen fibres and a more perpendicular arrangement of blood vessels (in relation to the overlying epidermis). Curiously, the vasculature within this area appeared to be exclusively comprised of blood vessels, with the endothelial cells showing positive labelling with CD31 but not D2-40 (Fig. 2E,F).

Lymphoedema can be primary or secondary in character. Primary lymphoedema is caused by inherited genetic defects resulting in impaired lymphangiogenesis. In contrast, acquired or secondary lymphoedema results from mechanical lymphatic failure due to trauma, infection or inflammation.[1] In developing countries, filariasis is the most common cause of secondary lymphoedema, while in Western countries the most commonly affected patient group are those undergoing treatment for malignancy (particularly breast or gynaecological malignancy) with lymphadenectomy and/or radiotherapy.[2] In recent times obesity has also been identified as both a cause and an aggravating factor for lymphoedema.[1] The cardinal histological features of lymphoedema comprise dermal oedema, fibroplasia, dilated lymphatic vessels and uniformly distributed spindle and dendritic cells.[1] Superimposed upon these changes may also be dermal inflammatory infiltrates and epidermal changes such as hyperkeratosis, acanthosis and papillomatosis. The clinical and histological

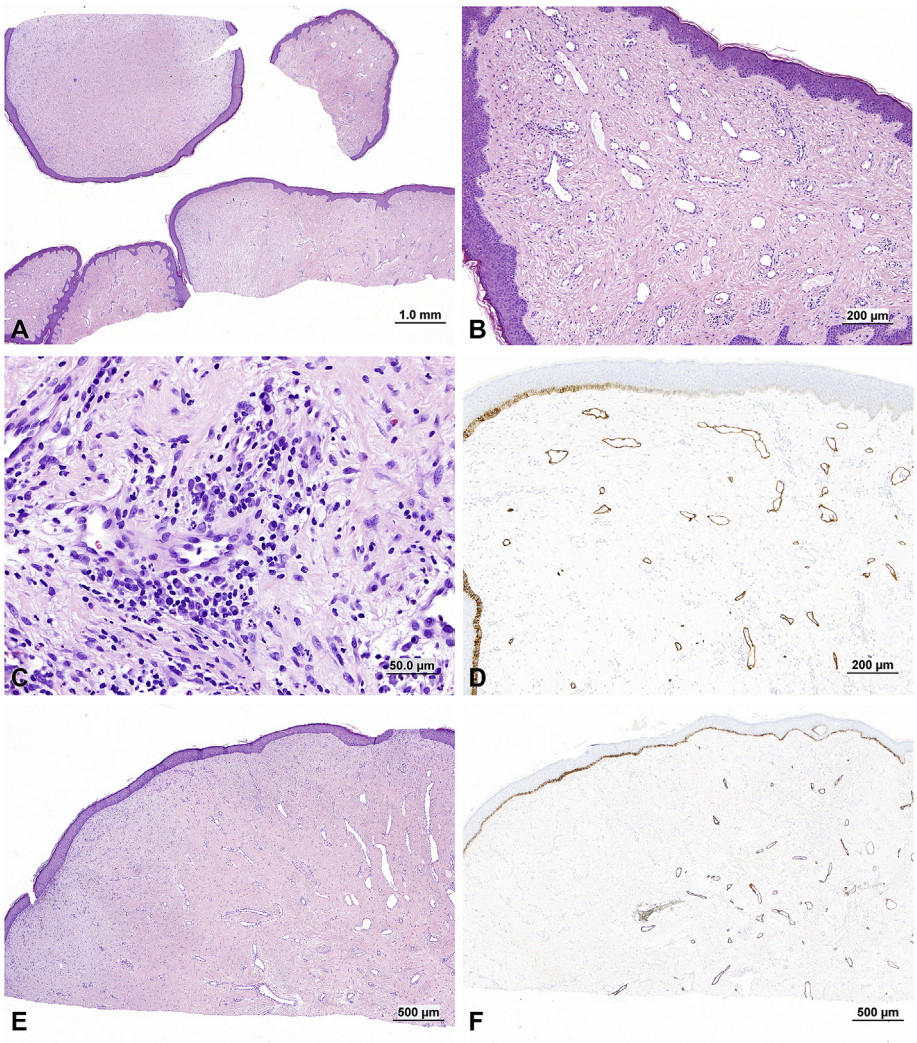

Fig. 2 The histological features of the lesion. (A) Low and (B) medium power views showing a papillomatous architecture, dermal fibrosis, dilated vasculature and evenly distributed spindled cells. (C) Foci of inflammation, including neutrophils and plasma cells, were also evident. (D) Immunohistochemical staing for D2-40 shows that many of the dilated vessels are lymphatic in nature. (E) An area of dermal scarring was also noted, subtly distinct from the lymphoedematous areas (scar to the left of the image). (F) No dilated lymphatics are seen in this area. (A, B, C, E: haematoxylin and eosin stain; D, F: immunohistochemical stains using antibodies against D2-40.)