

Nestle *et al.*<sup>7</sup> suggested that DF is an abortive immune process mediated by dermal dendritic cells, thus, any drug down-regulating T cells, such as imatinib in our cases, might favour the appearance of multiple DF through an exaggerated response to an unknown pathogen.<sup>2</sup> We found only one case report of MEDF in a patient with CML.<sup>8</sup> That case occurred after an aplastic episode and the authors considered the MEDF secondary to immunosuppression. Our second case could be included in this second group, as he presented the lesions during a worsening of his disease due to a failure of imatinib treatment, but our first patient would fit better taking into account the Nestle's hypotheses.

Our patients did not present any other sign of immunosuppression or other systemic diseases. This supports the pathogenic role of imatinib in the development of the MEDF. So, dermatologist should be aware of the appearance of MEDF in patients with CML as it should be an alarm bell to start a work up to rule out immunosuppression or treatment failure.

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DOI: 10.1111/jdv.12328

## Adult-onset eccrine angiomatous hamartoma: report of a rare entity with unusual histological features

Editor

Eccrine angiomatous hamartoma (EAH) is a rare hyperplasia of eccrine glands and dermal capillaries. It typically presents during childhood as a slowly growing, bluish-purple nodule or plaque on the distal extremities, and may be associated with pain, hyperhidrosis, and/or hypertrichosis.<sup>1</sup> We report a rare case of adult-onset EAH notable for its size and previously unreported histological features that could cause diagnostic confusion with inflammatory panniculitis.

Two years prior to presentation, a 44-year-old female developed a 3–4 cm, bruise-like lesion on the left thigh. She reported occasional pruritus, burning and 'tightness' in the lesion, but denied excessive sweating or hair growth. A punch biopsy performed by an outside practitioner was suggestive of morphea profunda. Despite treatment with topical steroids and calcipotriene, the lesion continued to grow, and initiation of methotrexate was being considered.

The patient presented to our dermatology clinic for a second opinion. Examination revealed a smooth 15 cm × 8 cm, irregularly shaped, mottled, red-brown plaque without hypertrichosis or hyperhidrosis (Fig. 1). Within the lesion, several soft, non-tender subcutaneous nodules were palpable.

To aid in diagnosis, we performed two 6-mm punch biopsies (Fig. 1). The first from a flesh-coloured nodule was non-diagnostic. The second biopsy from a red-brown nodule demonstrated hyperplastic dermal eccrine coils, prominent perieccrine mucin, numerous intimately associated capillaries, and a moderately dense, superficial and deep perivascular dermal lymphocytic infiltrate with occasional eosinophils (Fig. 2). Dermal collagen appeared normal. In the subcutis were prominent, widened, fibrotic septae containing multiple, small periseptal lymphocytic aggregates without follicle formation. Spirochete immunostain was negative.

The histological differential diagnosis included EAH with unusual histological features vs. a primary inflammatory panniculitis. Despite finding periseptal lymphoid aggregates, the presence of dermal eosinophils and the lack of a lobular lymphocytic panniculitis or hyaline sclerosis rendered lupus panniculitis unlikely. In addition, anti-nuclear antibody testing was negative. Although septal thickening was present, the degree of dermal inflammation and the clinical presentation of a persistent plaque on the thigh were not consistent with erythema nodosum. Lastly, the absence of hyalinized collagen excluded inflammatory morphea.



**Figure 1** Clinical presentation of eccrine angiomatous hamartoma (EAH). A 44-year-old female presented with a slowly growing, smooth, red-brown, mottled plaque on left medial thigh with flesh-coloured (biopsy site 1) and erythematous nodules (biopsy site 2).

Altogether, the clinical and histological features were most consistent with adult-onset EAH. The presence of striking dermal eccrine and vascular hyperplasia was pathognomonic. As the prominent dermal inflammation and subcutaneous septal fibrosis were not consistent with a specific type of inflammatory panniculitis, these changes were interpreted as incidental and secondary. The patient deferred excision due to lack of symptoms, and is currently doing well with observation.

Eccrine angiomatous hamartoma is a rare hamartoma that is typically solitary (86.5%) and located on the extremities (81.1%).<sup>1</sup> Symptoms may include pain (42.4%) and hyperhidrosis (34.3%).<sup>1</sup> In a review of 37 cases, 45.7% of lesions were congenital, with age of presentation ranging from 2 months to 73 years (median age 10 years).<sup>1</sup> Only 14 cases of adult-onset EAH have been described.<sup>2,3</sup> The reported sizes of EAH range from 3 mm to 11 cm.<sup>4</sup> Thus, our patient's lesion is the largest reported to date.

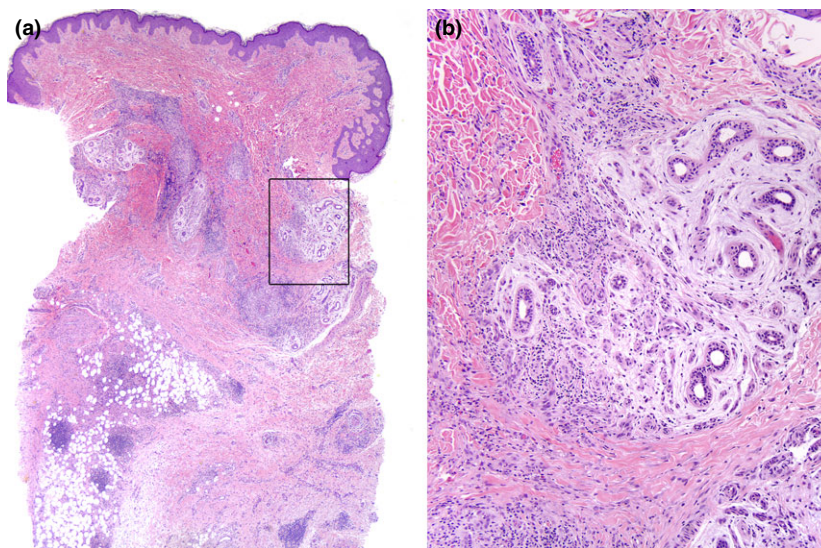
Both proliferation of eccrine glands and closely associated capillary angiomatous foci must be present to distinguish EAH from eccrine nevus and sudoriparous angioma, which lack capillary proliferation and eccrine hyperplasia, respectively.<sup>1</sup> Malignant transformation has not been reported.<sup>3,5</sup>

The treatment of choice for EAH is surgical excision and is required if there is pain, severe hyperhidrosis or unremitting growth.<sup>1</sup> Observation is reasonable for asymptomatic lesions.<sup>1,3</sup>

Awareness of the clinical and histological features of EAH is key to diagnosing this rare hamartoma, particularly in adults. Distinguishing EAH from inflammatory panniculitis will avoid aggressive or ineffective treatments. Our case demonstrates hallmark histological features of increased eccrine glands in intimate association with increased vessels. Inflammation and septal fibrosis are unique findings that have not been reported.

#### Acknowledgement

The authors thank Ms. Laura Van Goor, BFA, for assistance with images and figures.



**Figure 2** Eccrine angiomatous hamartoma (EAH) with unusual histological features. (a) Low-powered view showing hyperplastic eccrine coils in the mid-to-deep dermis with unusual features, including a dense perivascular inflammatory infiltrate and widening of pannicular septae (haematoxylin–eosin, original magnification  $\times 20$ ). (b) High-powered view of hyperplastic eccrine glands with perieccrine mucin and intimately associated increased small blood vessels (haematoxylin–eosin, original magnification  $\times 100$ ).

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DOI: 10.1111/jdv.12335