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Case Report

ECCRINE ANGIOMATOUS HYPERPLASIA – A RARE AND UNUSUAL MIMICKER OF NEUROFIBROMA

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Abstract
Eccrine angiomatous hyperplasia is a rare benign hamartomatous lesion categorized by proliferation of eccrine glands and small blood vessels. These lesions commonly involve the acral region appearing as papules, plaques or nodules and may be associated with hyperhiderosis and pain. We present a 32 year old male with massive bosselated swelling involving more than half of the lower limb, clinicoradiologically mimicking a neurofibroma.
Keywords: Eccrine angiomatous hyperplasia, hyperhiderosis, hamartoma, neurofibroma, histology.

Introduction

Eccrine angiomatous hyperplasia (EAH) is a type of benign hamartomatous cutaneous lesion characterized by proliferation of eccrine glands and vascular channels in the dermis. [1]. It is usually congenital and presents at birth or later in childhood presenting as single or multiple nodules, predominantly affecting the extremities. [2] There is paucity of literature regarding this rare and interesting entity. We report a case of this rare benign hamartoma in a 32 year old male who presented with a diffuse swelling over the right leg since six years of age.

Case Report A 32 year male presented to the plastic surgery department with a painless swelling of the right leg since six years of age. The lesion began at the ankle and gradually progressing upwards involving the knee and the lower third of the thigh. Sixteen years ago, he had undergone a debulking procedure in a tertiary centre and was diagnosed with neurofibroma. Following this, the swelling continued to increase in size. The patient had difficulty in walking and gave a history of profuse perspiration at the site of the lesion. Physical examination revealed a boss elated swelling of the lower limb extending to the thigh with superficial dilated veins. No surface ulcerations were seen. On palpation, multiple nodules were felt over the swelling. However, no pulsations were present owing to severe edema. There was bowing of the tibia and lateral displacement of the patella.

Systemic examination was unremarkable. On haemotological investigation he was detected to have thalassemia trait. Biochemical parameters were normal. Magnetic resonance imaging (MRI) of the right lower limb showed an ill defined heterogenus lesion with altered intensity, diffusely infiltrating the skeletal muscle. Proximally, the lesion was seen extending into the lower third of the thigh along the lateral aspect of the knee encasing the hamstring tendons and iliotibial tract and extending into the dorsum of foot inferiorly. Areas of interspersed fat and multiple dilated and tortuous blood vessels were also noted. There was patello-femoral joint dislocation with laterally dislocated patella. The possibility of a plexiform neurofibroma or hemangioma was suggested on radiology. A clinicoradiological diagnosis of neurofibroma was proffered. Surgical excision of the lesion was planned. However, due to excessive intraoperative bleed, the complete lesion could not be excised. The excised specimen was sent for histopathological examination. Grossly, the specimen was a single skin covered corrugated mass measuring 20x5x4 cm. Cut section showed grey brown solid areas and cystic spaces exuding serosanguinous fluid (Figure 1). Microscopy revealed a hyperkeratotic, focally acanthotic epidermis overlying dermis containing numerous thin and thick walled dilated vascular channels with splaying muscle wall, eccrine gland hyperplasia surrounded by a fibromyxomatous stroma with adipocytes proliferation (Figure 2). On immuno-histochemistry, few stromal cells and eccrine glands showed immunoreactivity for S100 (Figure 3) and the vessels demonstrated CD34 positivity (Figure 4). Based on clinical presentation, radiology and microscopy, a diagnosis of eccrine angiomatous hamartoma was rendered.

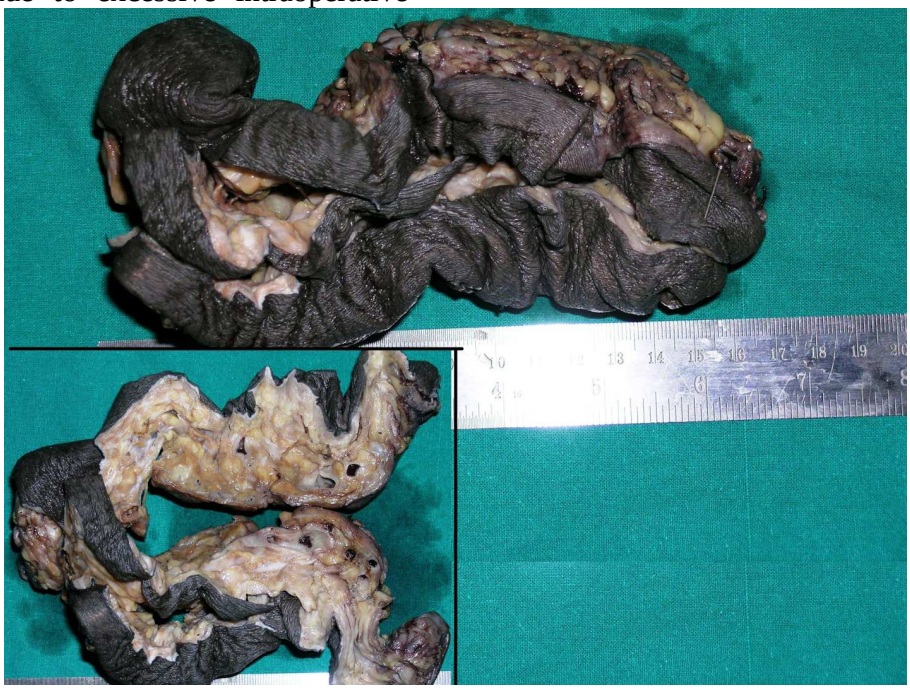

Figure 1: Excised specimen showing skin covered corrugated tissue mass with yellow fatty areas and cystic spaces on cut section (inset).

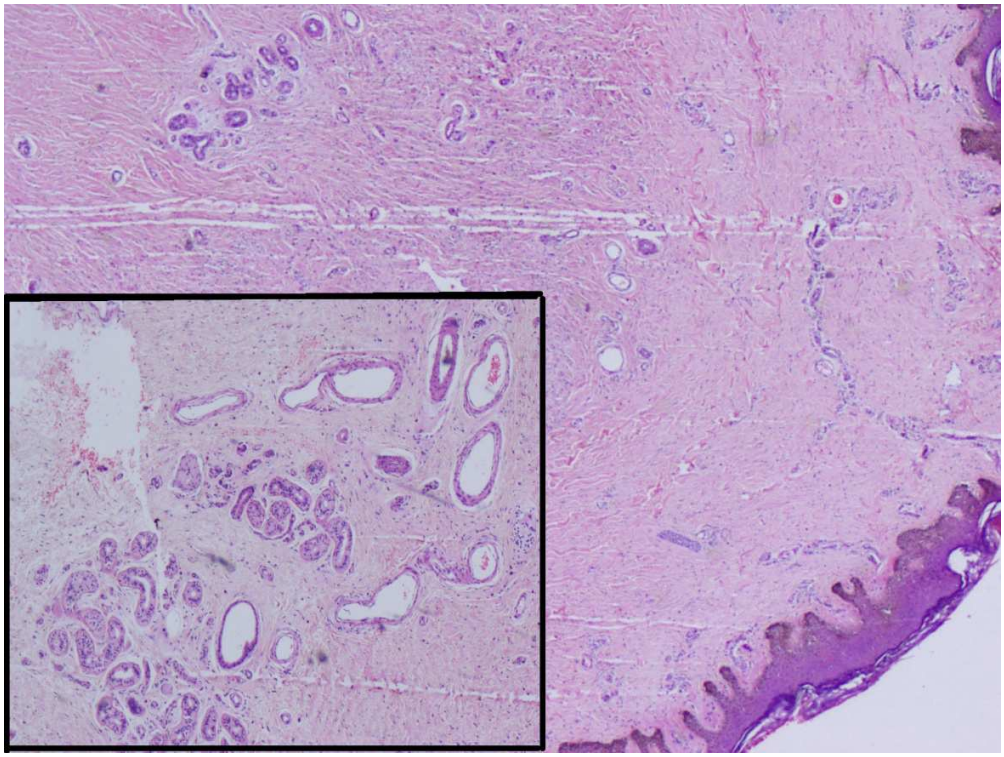

Figure 2: Photomicrograph showing hyperkeratotic epidermis overlying mid and deep dermis with (EAH, H&E, and 2 X). Inset shows hyperplasia of eccrine glands and proliferating medium and small sized blood vessels (H&E, 10X)

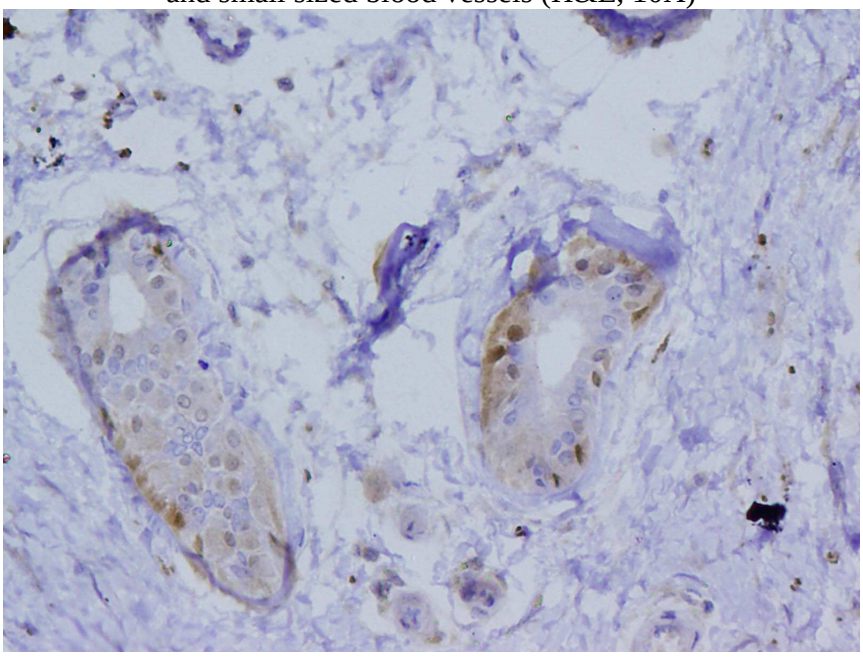

Figure 3: Secretory portion of eccrine positivity showing S100 positivity (20X)

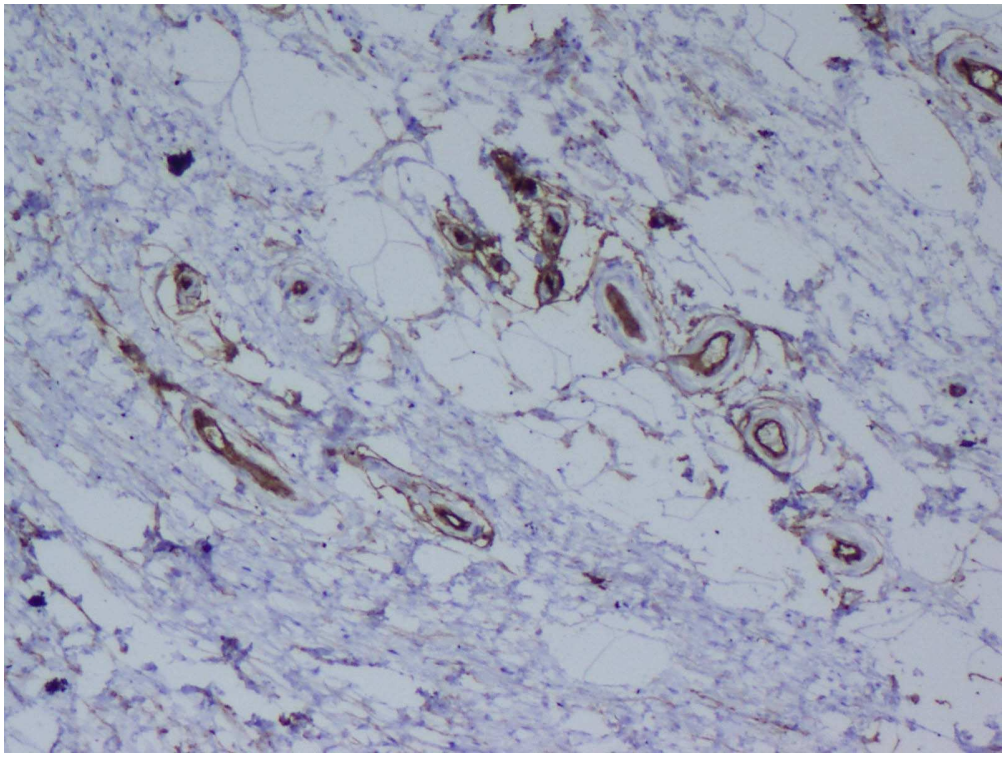

Figure 4: Proliferating blood vessels showing CD34 positivity (20X)

Discussion

EAH is a rare nevoid proliferation comprising predominantly of eccrine glands and small blood vessels. Adipose tissue and pillar structures may occasionally be present. [3] The pathogenesis of this malformation has been attributed to a defect in the interaction between differentiating epithelium and the surrounding connective tissue. [4]

EAH has no sex predilection and is mostly congenital. The lesion may also appear in early childhood as in our case. [5] Adult onset lesions have also been reported. [6,7] The common sites of involvement include palms and soles, and occasionally buttock, feet, face, neck and trunk. [1,2,3,8] They present as a gradually progressing solitary or multiple flesh colored, erythemous papule, plaque or nodule, measuring up to a few centimeters. These lesions are usually asymptomatic but may be associated with pain, hypertrichosis and hyperhidrosis. Rapid growth of the lesion has been documented during pregnancy and puberty owing to hormonal influences [1,2,5,9,10] The lesion in the present case was unusual in that it was massive involving the entire lower limb, causing difficulty in walking and associated with profuse sweating at that site. A similar case of EAH causing difficulty in walking due to painful plaque over the thigh has been reported. [10]

Ultrasonography aids in detecting the vascular nature of the lesion while MRI detects the exact size and relation with adjoining structures. [7]

The clinical differentials include neonatal angiomatosis, vascular malformations, tufted angioma, smooth muscle hamartoma, glomus tumor, blue rubber bleb nevus and macular telangiectatic mastocytosis. Radiologically, EAH may even mimic varicose veins at atypical sites. The presence

of associated symptoms such as pain, hypertrichosis and hyperhidersosis may be a useful clue to exclude the above lesions, however, definite diagnosis is made on histological examination of the lesion. [2,3,7] In our case, the clinical and radiological diagnosis was conclusive of neurofibroma which was excluded on histopathology.

Histologically, EAH is composed of well differentiated sweat glands along with angiomatous channels which are diagnostic. Fat infiltration, presence of mucin and other adnexal structures can also be present. [1] Immuno-histochemically, the eccrine glands show positivity for S100 and CEA while the vessels stain for antifactor VIII- related antigens and CD34. [1,3,5] EAH has to be differentiated from eccrine nevus by the lack of vascular channels in the latter. [1,5]

EAH is a slow growing lesion and simple excision is often curative. Botulinum toxin and intralesional sclerosants are newer treatment options for this lesion. [1,10] In the present case, the massive size of the lesion causing difficulty in walking warranted surgical excision of the lesion. However, due to intraoperative bleed, the lesion was not excised completely.

Conclusion

To conclude, EAH is an uncommon benign malformation with varied clinical presentation which is confirmed only on histopathology. Most often these tumours are indolent and can be treated by simple excision. The present case is different as it clinicoradiologically mimicked a neurofibroma and the giant size warranted aggressive surgical treatment.

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