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Case Report

Adult-onset eccrine angiomatous hamartoma treated with excision and skin grafting: a case report

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ABSTRACT

Eccrine angiomatous hamartoma is a rare benign tumour like malformation characterized by intradermal proliferation of eccrine and vascular components. It usually arises before puberty but can occur at any age. It typically presents as a solitary or multiple reddish or brownish plaque or nodule that progressively enlarges in size. Symptoms such as pain and hyperhidrosis may be associated. Lesions are commonly located over distal extremities. We report a case of a 45-year-old lady who presented with a gradually enlarging solitary reddish plaque over the left retro-auricular region of the scalp since the last 12 years and associated with occasional pain. Histopathology revealed pproliferation of eccrine sweat glands and thin-walled blood vessels in the dermis. Additional ultrasound and MRI contrast studies were done to assess the vascular component and involvement of underlying structures. The patient was treated successfully with complete excision of the lesion followed by split thickness skin grafting.

Keywords: Eccrine angiomatous hamartoma, Eccrine proliferation, Vascular proliferation

INTRODUCTION

Eccrine angiomatous hamartoma is an uncommon hamartomatous tumour characterised by proliferation of eccrine and vascular structures in the dermis. It generally arises at birth or later in childhood. Adult-onset cases have also been reported. It commonly presents as a single nodule or papule but can present with multiple lesions occasionally. The lesion may be associated with pain, hypertrichosis or hyperhidrosis which raise a suspicion of EAH, but histological confirmation is required for final diagnosis.[1]

CASE REPORT

A 45-year-old lady came to dermatology OPD with complaints of single raised lesion over left retro auricular area for 12 years. It started as a small pea sized swelling and gradually increased to present size over a period of 12 years. It was associated with on and off pain for 6 months. There was no history of trauma/surgery prior to the onset of lesion. No history of bleeding or discharge from the lesion. No history of hyperhidrosis over the lesion. Her past medical history was not significant. No history of pulmonary tuberculosis, hypertension, diabetes mellitus, bronchial asthma or seizures.

Cutaneous examination showed a solitary, erythematous, tender plaque, approximately measuring 6x4 cm in size present over the left retro-auricular area (Figure 1). Its margins were well defined, consistency was soft and surface was lobulated. Hypertrichosis and hyperhidrosis were absent. Bruit was not heard over the lesion.

Ultrasound revealed an iso to hypoechoic soft tissue lesion measuring 6 mm in its maximum thickness

involving cutaneous and subcutaneous plane of left post auricular region with no evidence of calcification, extension into deeper planes or underlying bony cortical irregularity and on colour doppler showed minimal vascularity. Since lesion was present on the scalp, MRI brain was performed. MRI revealed a well-defined T1 isointense and T2/flair hyperintense signal lesion measuring approximately 3.1x2.8x4.5 (APxTRxCC) involving subcutaneous plane of left mastoid region. There was no evidence of calcification/fat component. Post contrast study showed significant enhancement. There was no evidence of diffusion restriction. Thus, intracranial involvement was ruled out.

Figure 1: Solitary, erythematous, tender plaque over the left retro-auricular area.

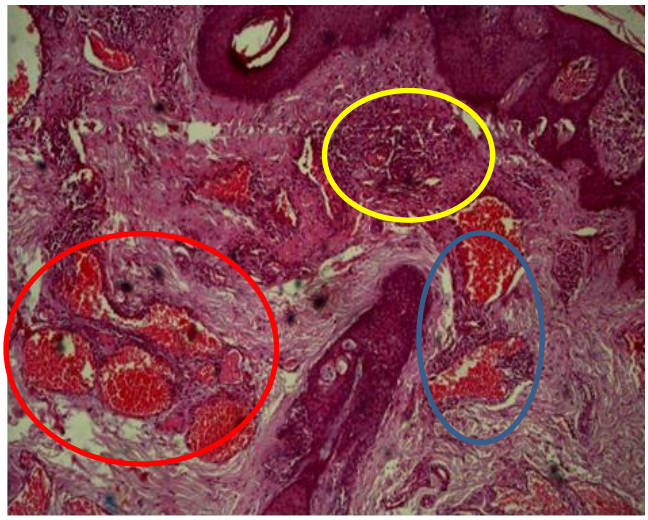

Figure 2: Photomicrograph of histopathology of proliferation of eccrine sweat glands and blood vessels. Dilated blood vessels seen adjacent to the eccrine glands and arranged in clusters. Proliferation of eccrine glands (yellow circle), dilated thin-walled blood vessels adjacent to the eccrine glands (Blue circle), cluster of blood vessels with adjacent eccrine glands (red circle).

Histopathological examination revealed parakeratosis, granulosis, acanthosis and elongation of rete ridges in epidermis. Proliferation of eccrine sweat glands and blood vessels which were thin walled, dilated and filled with RBCs were seen in dermis and subcutaneous tissue. The blood vessels were seen adjacent to the eccrine glands and were arranged in clusters. Superficial dermis showed lymphocytes and plasma cell infiltrate (Figure 2).

Since the lesion was large and involving the scalp, patient was referred to plastic surgery department, who performed excision of the tumor with split thickness skin grafting.

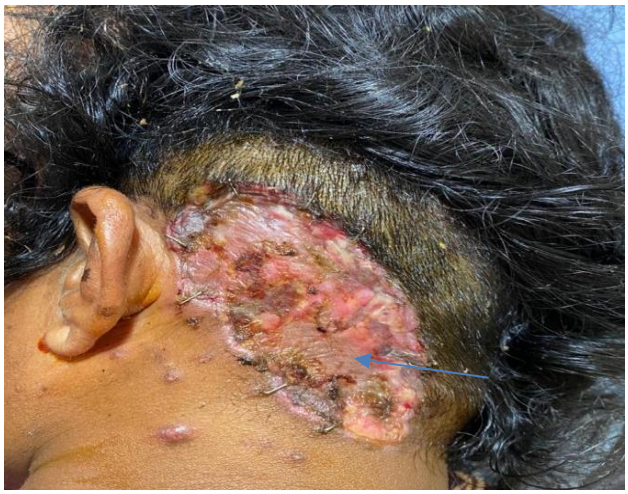

Figure 3: Split thickness skin graft following excision of the tumour.

DISCUSSION

The term eccrine angiomatous hamartoma was coined by Hyman et al in 1968. However, in 1859 a similar case was reported by Lotzbeck et al.[2] Eccrine angiomatous hamartoma also known as Eccrine angiomatous nevus is a rare benign tumour of hamartomatous nature, characterised histologically by proliferation of eccrine secretory coils and ducts with surrounding capillary angiomatous channels and occasionally adipose tissue, mucin and neural elements.

This disease usually presents during childhood and adolescence and shows a male: female ratio of 1.2:1.[3] Almost 70% of EAH arise before puberty (approximately 48.9% are congenital lesions, while 23.4% appear in late childhood), but patients presenting in adulthood have also been described.[4]

The exact pathogenesis of EAH is unknown, but various theories such as abnormal induction of heterotypic dependency with resultant malformation of adnexal as well as mesenchymal elements have been proposed.[5] The mechanisms of multiple EAH spreading to several parts of the body may be due to mosaicism of gene mutation occurring in early developmental stage. It has also been associated with radiation therapy and repeated trauma.[3] It occurs due to abnormal biochemical interaction between

epithelium and mesenchyme resulting in atypical proliferation of eccrine and vascular elements.

The tumour usually presents as a solitary nodule or papule that may be flesh coloured or bluish or violaceous or red. The most common site of predilection is distal extremities. The lesion may also be associated with hypertrichosis or hyperhidrosis or pain which aids in the diagnosis of EAH.

Histopathologically, it is characterised by diverse features like proliferation of vascular and eccrine components, presence of mucinous, lipomatous and/or lymphatic structures in dermis and subcutaneous tissue. Pella et al proposed a diagnostic criterion for EAH which includes- a) Hyperplasia or normal or dilated eccrine glands, b) Close association of eccrine structures with capillary angiomatous foci and c) The variable presence of pilar, lipomatous, mucinous and/or lymphatic structures. Smith et al also described three histological variants of EAH - Follicular, lipomatous and mucinous. Immunohistochemistry has been carried out only in few cases of EAH. Immunohistological pattern of EAH is similar to that of normal eccrine gland and capillary channels. Eccrine sweat apparatuses stain positive for S100, carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), and CAM5.2 antibody. Vascular components show positivity for CD34, CD44, human nerve growth factor receptor and *Ulex europaeus* and factor VIII antigen. The surrounding stroma show positivity for anti-CD36.[3,6]

In one of the cases described by Sanusi et al, ultrasonography showed diffuse lesions with increased blood flow and MRI revealed contrast enhanced subcutaneous lesion with diffuse signal intensities in T1 and T2 weighted sequences.[3]

Differential diagnosis for EAH include vascular malformations, tufted angiomas, smooth muscle hamartomas, glomus tumour, blue rubber bleb naevus, macular telangiectatic mastocytosis, eccrine nevus and sudoriparous angioma.[8,9]

EAH does not commonly require treatment due to its benign nature. Malignant transformation has not been reported till date.[7] Spontaneous regression has been reported by Tay and Sim in one patient.[3] Medical intervention is generally indicated either for pain or for cosmetic reasons. The main line of treatment is surgical excision.[10] Localized and smaller lesions can be treated by simple local excision. Larger or painful nodules or plaques however may sometimes require amputation of involved digit or extremity to control the symptoms in those with larger lesions in the acral part. In cases where surgical removal poses practical difficulty, other options such as intralesional sclerosants, pulsed dye laser, Nd: YAG Laser and botulinum toxin can be used. Aethoxysclerol, which acts by causing damage to endothelium by attacking the lipid of the membrane, leading to cell damage and inflammation.[11] Hyperhidrosis and excessive neuromuscular activity are main indications for Botulinum toxin treatment in EAH. It is mainly used to suppress excessive sweat production of hyperplastic eccrine sweat glands in hamartoma. Barco et al successfully treated a 12-year-old girl with EAH with Botulinum toxin. The minor starch-iodine test was performed to assess the area to be treated and 2.5 U per injection of Botulinum toxin type A infused in a single course of 14 injections spaced 1.5 cm apart. Botulinum toxin has to be diluted with isotonic sodium chloride solution to a concentration of 2.5U/0.1 ml. Using an insulin syringe (1 ml), a total dose of 35 U was infused.[6]

CONCLUSION

Eccrine angiomatous hamartoma is an uncommon entity. Lesions usually occur in the extremities. However, in our patient the lesion was on the scalp which is an uncommon site of involvement. The lesion was successfully excised and skin grafting was done. Hence, we report this case for its rarity and peculiar site of involvement.

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