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Eccrine Angiomatous Hamartoma- A Rare Clinical and Histopathological Presentation

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Abstract
Background: *Eccrine Angiomatous Hamartoma is a vascular lesion of the skin. It is a rare benign tumor characterized by proliferation of eccrine and vascular components.*
Case Details: *We present an interesting case of 7 year old male who had a plaque like tumor present since birth over the heel of left leg which was gradually progressing to form a nodular swelling histopathological examination consist of proliferation of hyperplastic eccrine glands with dilated capillaries .the overlying epidermis showed marked verrucous changes which are reported in only a few cases of Eccrine Angiomatous Hamartoma .*
Conclusion: *Eccrine Angiomatous Hamartoma is a rare, slow growing benign tumor characterized by increased eccrine structures and capillaries. Epidermal change can be mistaken for verrucous skin lesion. Recognition of the lesion is important to decide on surgical intervention as simple excision is curative for a painful and aggressive tumor only.*
Keywords: *hyperkeratosis, parakeratosis , eccrine glands, verrucous , hamartoma.*

Introduction

Eccrine Angiomatous Hamartoma (EAH) is a vascular lesion of the skin which was first coined in 1968 by Hyam et al but was described in 1859 by Lotzbeck.[1-5] It is a rare benign tumor characterized by proliferation of eccrine and vascular components[1,2]. It usually presents at birth in about 50% of patients, in 25% it appears later in childhood, adult cases have also been reported[6]. We present this case of Eccrine Angiomatous Hamartoma in a 7 year old male for atypical clinical and histopathological findings.

Case Summary

A 7 year old male presented to our Out Patient surgery Department with a complaint of painful swelling over left heel. It was present since birth and had progressively enlarged with age. He had never consulted in past for any skin conditions. There was no history of trauma, no history of any systemic problem or family history of similar lesion. Physical examination revealed a well circumscribed nodule with thickened overlying skin, measuring 1.5x 0.5 cms. The adjacent skin was brownish in color surrounding the nodule measuring around 1.5 cms. There was no evidence

of hypertrichosis or hyperhydrosis. Clinical diagnosis of Callus, fibromatosis, Foreign body granuloma, fibroma, neurofibroma was considered. Fine needle aspiration cytology revealed scanty cellularity with few fragments of adipose tissue, few clusters of round to oval cells, spindle shaped cells against hemmoraghic background. No conclusive diagnosis was possible on FNA. Surgical excision was done which revealed an ill circumscribed fibroelastic tissue. Histopathology examination (figure 1) revealed epidermal changes like hyperkeratosis, parakeratosis, acanthosis and papillomatosis, the deep dermis revealed a well circumscribed unencapsulated nodule. The nodule was composed of eccrine glands admixed with small and large blood vessels in a fibrofatty connective tissue. No evidence of atypia was seen. The histopathology was consistent with the diagnosis of Eccrine Angiomatous Hamartoma.

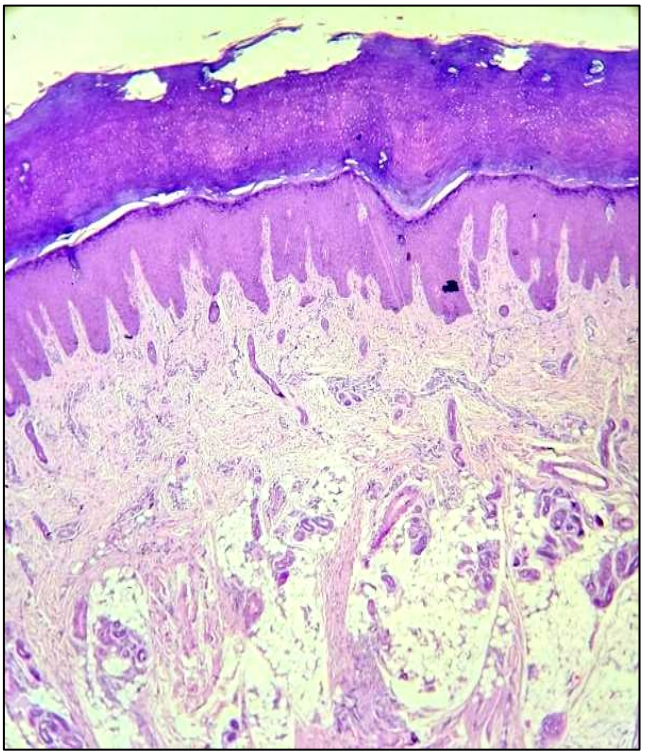

Figure (1) In Epidermal changes like hyperkeratosis, parakeratosis, acanthosis and papillomatosis seen, the deep dermis revealed a well circumscribed unencapsulated nodule . The nodule was composed of eccrine glands admixed with small and large blood vessels in a fibro fatty connective tissue. (40x magnification)

Discussion

Eccrine Angiomatous Hamartoma is a rare benign cutaneous tumor usually present at birth or early infancy or childhood. However there have been cases reported even in adults[7]. It can occur as a result of abnormal interaction between differentiated epithelium and underlying mesenchyma which leads to atypical proliferation of eccrine and vascular elements[8] True incidence is yet unknown[2,4] and there is no gender preponderance[2,3,4]. It manifests as a solitary lesion [1,2,4] which affects limbs and more particularly the plamoplantar areas [9], Our case was located on the heel. Other sites which are affected are trunk and neck[1,5,10.] Rare sites like eyelids, cheeks, vulva have been reported[11]

It has varied presentation and can appears as plaques, nodules and uncommonly as papules and macules[1,2]. Color of the lesion may be violet brown, blue .red or pink. It can also be of normal skin color. Martinelli and Zeller[8],[12] have described atypical variants with superficial changes resembling linear verrucous lesion, acanthosis nigrans and hypertrichosis in their reports. Our case presented with verrucous lesion,. Some lesions have hyperhydrosis and hypertrichosis which are important clues for diagnosis of EAH. Most patients are asymptomatic but some may experience pain due to involvement of small nerves that penetrate the tissue[1,2.] Hormonal influence may play a role in causing pain and rapid growth of lesion as seen in puberty and pregnancy[1,2,9]. Our case presented with a painful nodule without hyperhydrosis or hypertrichosis.

Very few reports are available on the utility of Fine needle aspiration cytology (FNAC) for diagnosis of EAH. In our case the cellularity was scanty and had mixed cells of adipocytes, spindle shaped cells and round to oval cells which was not diagnostic.

Definitive diagnosis of EAH is by histopathological examination[2]. The hallmark is proliferation of eccrine and vascular structures consisting mainly of capillaries .Histopathologic variants have been reported which include

increased dermal mucin and infiltration in the adipose tissue. In our case adipose tissue element was found with eccrine glands, proliferating blood vessels with nerve fragments.

Pele et al proposed criteria for diagnosis of EAH[13].

1. Hyperplasia of normal or dilated eccrine glands
2. Close association of eccrine structures with capillary angiomatous foci and
3. Variable presence of pilar, lipomatous , mucinous and/or lymphatic structures

Immunohistopathological study was not performed in our case but studies report that CEA and S- 100 protein are reduced in Eccrine glands of EAH[14].

Our case showed marked verrucous changes in the epidermis which are reported in only a few cases of EAH and verrucous hemangioma like features [15,16]

The Differential Diagnosis is broad because of wide spectrum of clinical variants of this disorder . Differential Diagnosis should include angioleiomyoma, eccrine nevus, vascular lesion , childrens fibrous hamartoma, smooth muscle hamartoma.[1,4, 10,17] All these conditions are histopathologically distinguishable[17] .

EAH has a benign course and does not regress spontaneously or have a malignant transformation. Recurrence may occur. The surgical excision offers therapeutic management for small lesion and larger lesions may need aggressive surgeries. Surgery is indicated in patients who have intense pain, excessive sweating or for cosmesis[10,18]

Conclusion

Eccrine Angiomatous Hamartoma is a rare, slow growing benign tumor characterized by increased eccrine structures and capillaries. Recognition of the lesion is important as simple excision is curative for a painful and aggressive tumor only .We report the case for rarity of site that is the heel, presence of unusual epidermal changes which can be mistaken for verrucous skin lesion and presence of adipose tissue as an unusual histopathological variant in an Eccrine Angiomatous Hamartoma .

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