CASE REPORT

ECCRINE ANGIOMATOUS HYPERPLASIA WITH UNUSUAL FEATURES.

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ABSTRACT
Eccrine Angiomatous Hyperplasia (EAH) describes a rare angiomatous skin lesion which on histopathology shows prominent blood vessels and aggregates of eccrine glands. EAH is both clinically and histologically heterogeneous showing varied clinical and microscopic features. We report a case of 30 years old male presenting with a large linear plaque and episodes of spontaneous bleeding from the lesion. Histologically, in addition to the usual features of EAH there was also proliferation of sebaceous glands and areas reminiscent of nevus sebaceous. The presence of sebaceous differentiation suggests that these hamartomas may be of apocrine origin. The areas reminiscent of nevus sebaceous, warrants careful follow up for malignant transformation in our patient. To the best of our knowledge these features have not been documented before in EAH.
Keywords: *Eccrine Angiomatous Hyperplasia, Sebaceous differentiation*
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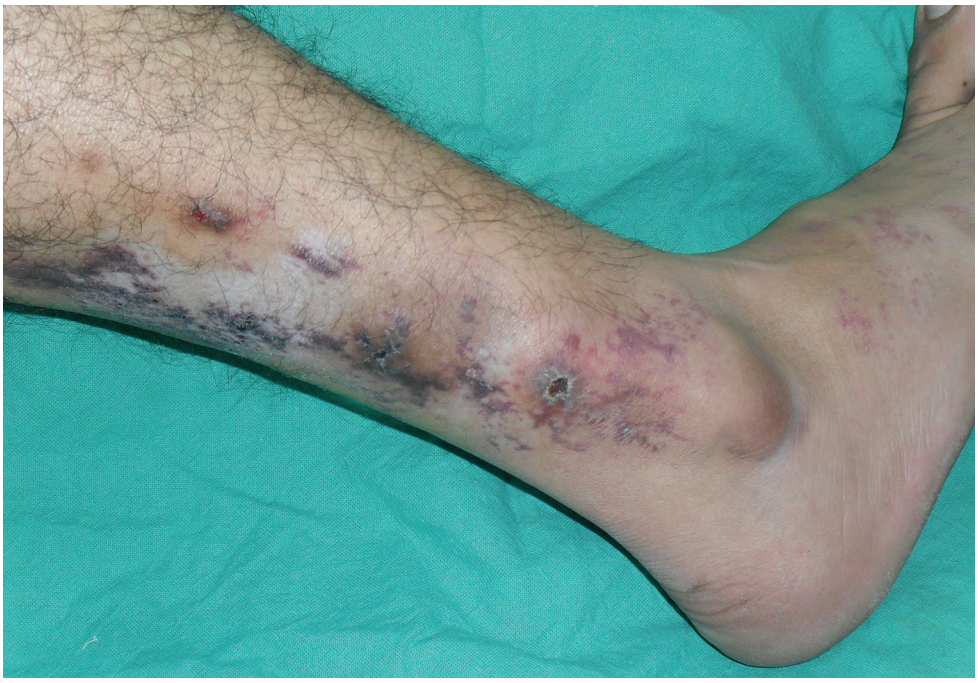

Figure 1: Clinical picture

INTRODUCTION

Lotzbeck [1] in 1859 described an angiomatous lesion on cheek of a child which on histopathology showed prominent blood vessels and aggregates of eccrine glands. The term Eccrine Angiomatous hyperplasia (EAH) was coined for such lesions by Hyman [2] in 1968. Since then less than 80 cases have been described in literature documenting proliferation of additional mesenchymal elements. A biochemical fault in the interactions between differentiating epithelium and subjacent mesenchyme that gives rise to an abnormal proliferation of mesenchymal, adenexal and vascular structures [3-5] has been proposed to explain the pathogenesis of EAH.

It usually presents as flesh colored, hyperhidrotic, hypertrichotic, painful macule, papule, nodule or a plaque on the extremities present congenitally or developing later on. We

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report a case of this rare entity in a 30 years old male with new additional clinical and histological features. Our patient had a large linear lesion with episodes of spontaneous bleeding from the lesion. Histologically, in addition to the usual features there was also proliferation of sebaceous glands and areas reminiscent of nevus sebaceous. To the best of our knowledge these features have not been documented before.

CAPSULE SUMMARY

The author reports a case of Eccrine Angiomatous hyperplasia (EAH) with the unusual features of proliferation of sebaceous glands and areas reminiscent of nevus sebaceous for the first time in literature. These new findings stress on the need of long term follow-up for development of local malignancy.

CASE REPORT

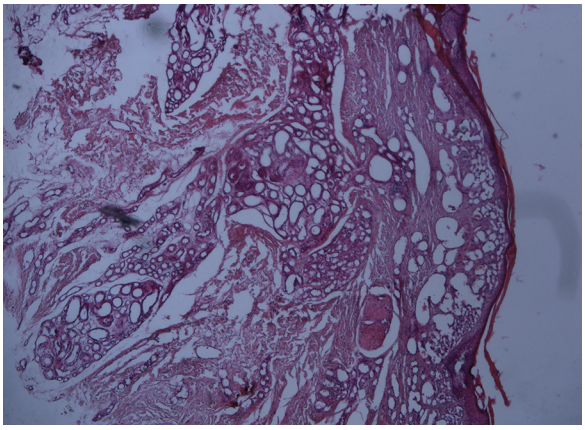

Figure 2: Histopathology

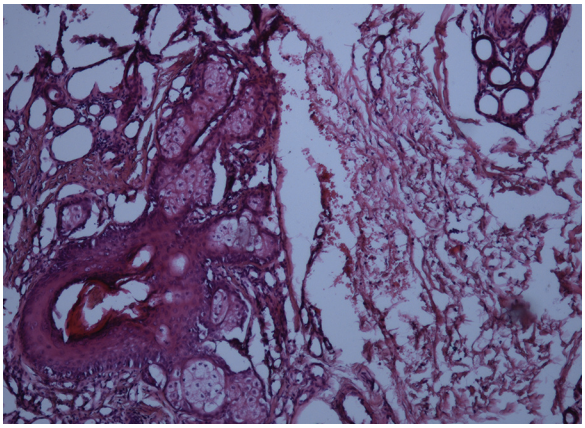

Figure 3a: Histopathology

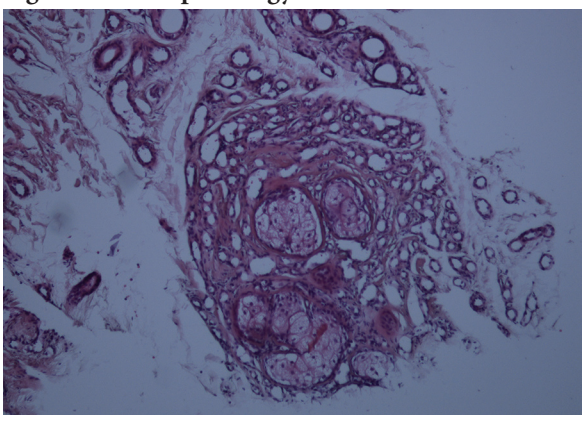

Figure 3b: Histopathology

A 30 years old male Pakistani presented with a 10 years history of a linear plaque over outer side of right leg. The plaque gradually increased in size over five years to involve the lateral side of the leg from mid calf to the planter and dorsal aspect of the right foot. The lesion was symptomatic in bleeding profusely on trauma and sometimes spontaneously. It was mildly tender and painful after minor trauma. There was no history of excessive sweating or hair growth over the lesion. There was no family history of similar lesions. On examination, a patchy erythmatous macular lesion was seen extending in a linear fashion from middle of right lateral calf to the planter and dorsal aspect of the right foot up to the bases of toes. The lesion measured 41 X 6 cm in its largest dimensions. At places the lesion was raised forming plaques with superficial erosions and crusts (Figure 1). At this stage a differential diagnosis of haemangioma and angiokeratoma was considered and skin biopsy was taken. Histopathology showed mild hyperkeratosis with normal prickle cell layer. There was marked dilatation and proliferation of thin walled blood vessels immediately below the epidermis causing compression of the overlying epidermis, reducing it to a thin fringe at places. There was widespread proliferation and dilatation of eccrine structures (Figure 2). At places these two structures intermingled with each other. There were scattered areas of dysgenesis of pilosebaceous units resembling nevus sebaceous (Figure 3a). At other places there was proliferation of sebaceous glands intermingled with eccrine structures (Figure 3b). On the basis of these findings, a diagnosis of EAH was made. The patient was briefed about the prognosis of the condition and was advised to avoid trauma in order to minimize bleeding from the lesion. Regular follow up was advised.

DISCUSSION

EAH is clinically and histologically heterogeneous, with a wide range of clinical and microscopic characteristics. Almost half of all lesions are present at birth, one-quarter appear in childhood, and the remainder appear in adulthood[2-5]. The most common clinical presentation is an acrally located papule, plaque, or nodule, though lesions on the trunk (25.5%) and scalp (2.1%) may also be seen [2-4]. In approximately 25% of cases, multiple lesions are present. The largest reported size is 8X11 cm [6]. In our patient the onset was in adulthood. The lesions were multiple angiomatous macules, nodules and plaques which were disposed on right lower leg and foot in a linear distribution and measured 41X6 cm in largest dimensions. Linear lesions in EAH have not been documented before. A case of extensive linear verrucous epidermal nevus showing mixed histological changes of verrucous epidermal nevus, nevus sebaceous, and eccrine angiomatous hamartoma has been described [7] but the lesions were clinically those of verrucous epidermal nevus in contrast to the angiomatous appearance as in our case.

Clinically Linear angiomatous arrangement caused confusion with segmental angioma and angiokeratoma in our patient.

EAH can be asymptomatic, painful (42%), or hyperhidrotic (32%) [8]. In our patient, the lesion was not hyperhidrotic and displayed mild tenderness which improved after partial excision. Our patient displayed spontaneous hemorrhage from the lesions, a feature not documented before. The most likely cause could be marked proliferation of blood vessels seen just underneath the epidermis causing it to be thinned to a small fringe and making it vulnerable to rupture with minor unnoticed trauma.

Histologically, EAH displays proliferation of normal or dilated eccrine glands, in close association with angiomatous foci and variable presence of pilar[9], lipomatous[10], mucinous[11], bony[12] and lymphatic structures[13] making it an organoid hamartoma. Acekerman reclassified many eccrine tumours as apocrine, and many adenexal that were previously thought to only show eccrine ductal differentiation now show ductal apocrine differentiation. Our patient had areas of dysgenesis of pilosebaceous units similar to nevus sebaceous, as well as sebaceous element proliferation, a previously unknown feature. Because of the close relationship between the apocrine and pilosebaceous units, our case could be an apocrine-folliculo-sebaceous unit hamartoma, indicating apocrine-sebaceous angiomatous hyperplasia.

Furthermore, epidermal organoid naevi with sebaceous elements have been linked to the development of a variety of benign and malignant neoplasms, possibly due to a loss of heterozygozity in the region of the human homologue of the Drosophila patched (PTCH) gene15.Although there was no evidence of malignancy in our patient, it should be carefully followed up in the above pretext.

In conclusion, our case has widened the clinical and histological spectrum of EAH by displaying very large linear angiomatous lesion showing spontaneous hemorrhage from the lesion. The presence of sebaceous elements histologically, argues in favour of the possible apocrine origin of these tumors as well as warrants careful follow up for malignant transformation in our patient.

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