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Eccrine angiomatous hamartoma in afour year oldgirlwith an illusion of angio-osteohypertrophysyndrome – A case report

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Abstract: Eccrine angiomatous hamartoma is a benign cutaneous malformation with unknown etiology. It usually originates from an abnormal interaction between the differentiated epithelium and the mesenchyme which results in atypical development of eccrine and vascular elements.It is usually asymptomatic, although focal hyperhidrosis, hypertrichosis, and pain may be observed. We report a case of this rare entity presenting in a four year old girl.

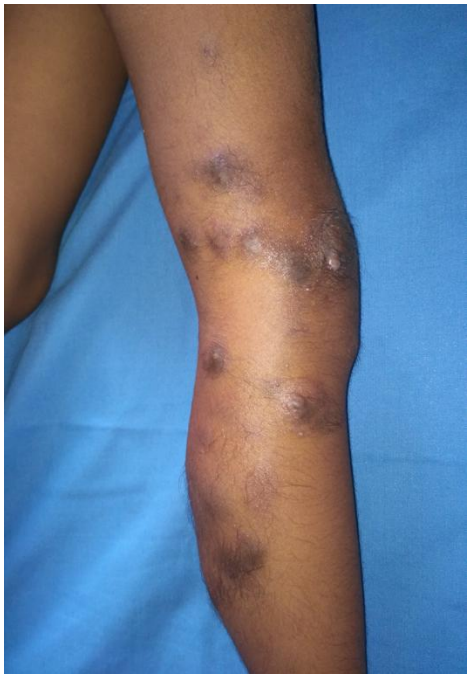
Keywords : Hamartoma , angio-osteohypertrophy syndrome.

Introduction :

Eccrine angiomatous hamartoma (EAH) is a benign and rare cutaneous tumor, histologically characterized by increased numberof eccrine sweat glands and numerous capillary channels[1].The term eccrine angiomatous hamartoma was coined by Hyman et al. in the year 1968[2]; however, the clinical description was first put forward by Lotzbeck, in 1895 and the earlier name was sudoriparous angioma.[3] Eccrine angiomatous hamartoma is usually present at birth or develops during childhood[4]. It is typically a solitary lesion and the most common site is on the distal parts of limbs.

Case Report:

A four year old girl presented to our op with multiple dark raisedlesions over the right leg which were present since birth and increased in size of lesions and girth of limb, with age. Initially the lesions were asymptomatic and later she developed occasional mild pain and increased sweating, hair growth here and there over the lesion. On examination multiple, discrete hyperpigmented firm plaques are present over the right leg associated with hyperhidrosis ,hypertrichosisand hypertrophy of the right limb[figure 1,2] without discrimination in the length of the limb, hence we came to differential diagnosisof angioosteohypertrophy syndrome and eccrine angiomatous hamartoma.Biopsy features are consistent with eccrine angiomatous hamartoma.[Figure 3,4].



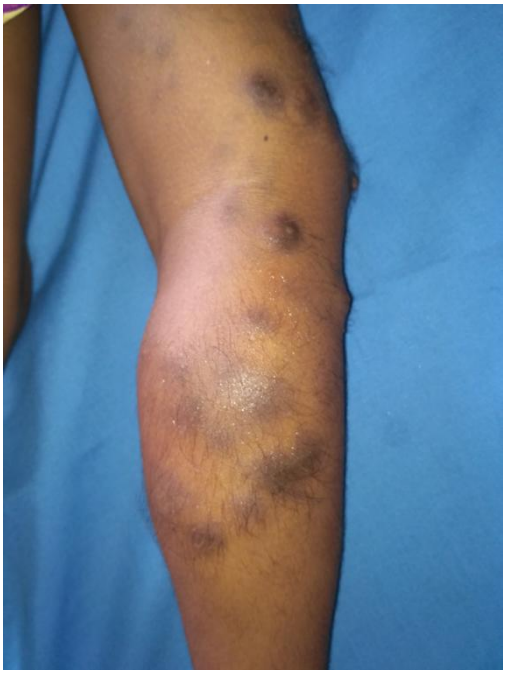
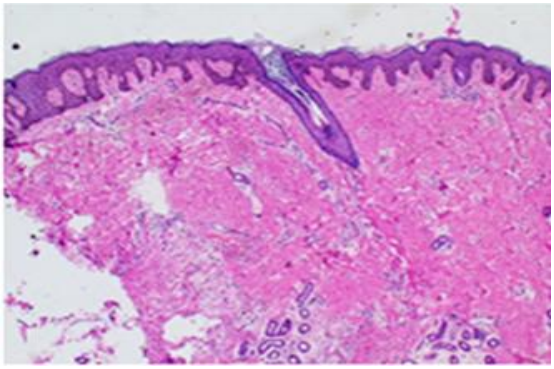


FIGURE 1 : multiple well defined dark brown plaques with the largest lesion measuring about 2 x 3 cm size are present over the posterior aspect of the knee extending onto the posterior aspect of the right leg.

FIGURE 2 : hyperhidrosis and hypertrichosis is seen over the lesions present present over the posterior aspect of the right leg.



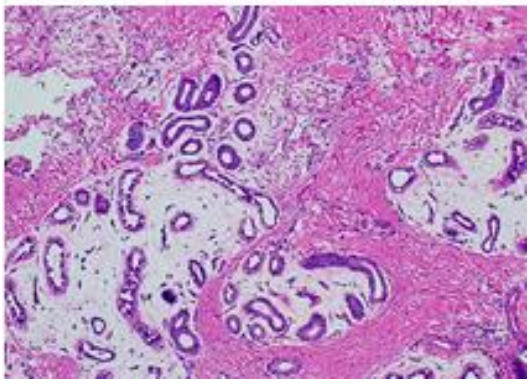


FIGURE 3 :Biopsy showed numerous thin walled dilated and collapsed fine venules throughout the dermis.

FIGURE 4 : Eccrine units are prominent and increased in number and are in close association with venules.

Discussion:

Eccrine angiomatous hamartoma is caused by proliferation of three appendages: eccrine glands, capillary channels and hair.Extremities, palms and soles in particular, are the usual sites affected[5]. Solitary, or multiple flesh-colored, blue-brown or reddish papules, plaques and nodules are characteristic. The criteria for the

diagnosis of EAH were proposed by Pelle et al. [6]as follow: (1) hyperplasia of normal or dilated eccrine glands, (2) close association of the eccrine structures with capillary angiomatous foci, and (3) variable presence of pilar, lipomatous,mucinous and/or lymphatic structures. Differential diagnosis include tufted angioma, eccrine nevus,vascular malformations, smooth muscle hamartoma ,blue rubber bleb nevus[7].The natural history of this tumor is benign and typically slow-growing and hence, aggressive treatment is generally unwarranted.Simple excision usually is curative and reserved for painful or cosmetically unacceptable lesions.[8]

Conclusion :

This case is reported due to its rarity.

Conflicts of interest :none

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