Congenital Eccrine Angio Keratomatous Hamartoma

Swasti Shubham, Upasana Uniya, Tina Rai, R A Hazari
Department of Pathology, People’s College of Medical Sciences & Research Centre, Bhopal-462037
(Received: January, 2017) (Accepted: January, 2017)

ABSTRACT

Eccrine Angio Keratomatous Hamartoma (EAKH) is a recently recognized skin lesion. It comprises of histological features of both Angiokeratoma and Eccrine Angiomatous Hamartoma (EAH). Only two cases have been reported in literature so far, both in adults. We are reporting the first case in a child. As a new entity, its awareness is required. Also, in case of a child it needs to be distinguished from other vascular lesions presenting in childhood.

KEY WORDS: angiokeratoma; eccrine; hamartoma; vascular malformation

INTRODUCTION:

Eccrine Angio Keratomatous Hamartoma (EAKH) is a skin lesion comprising of histological features of both Angiokeratoma and Eccrine Angiomatous Hamartoma (EAH). It was first reported as a distinct entity in 2006[1]. EAH is a rare tumor [1] comprising of benign dermal proliferation of eccrine sweat glands and capillaries[2]. Angiokeratomas are considered to be true ectasias of blood vessels in the superficial dermis [3]. After its recognition as a distinct entity, only one case of EAKH has been reported so far[4]. We are reporting the third case of EAKH.

CASE REPORT:

A one year old female child presented with black coloured papular lesions over left leg. The lesion was present since birth and increased in size over time. The lesions were three in number, the largest measuring 3 cm *1.5 cm. An ulcer with crusting was also noted in the largest lesion. The child did not have any difficulty in moving the leg. General and systemic examination did not reveal any abnormality. Routine hemogram and biochemical investigations were within normal limits. USG of the lesion showed prominent vascularity.

A punch biopsy of one of the lesions was done.

Grossly, the biopsy measured 0.8 cm. Histopathological examination of the biopsy showed hyperplastic and papillary epithelium with mild spongiosis. The papillary dermis was unremarkable. The deeper dermis and part of subcutis showed proliferation of numerous capillary channels intermingled with several eccrine glands. Fibrofatty tissue and pilar structures were also seen admixed. A diagnosis of EAH was made.

Following the biopsy report, complete excision of the lesions was done. The specimen received were three elliptical skin covered masses measuring 2.5 cm*1.7 cm*1 cm, 2 cm* 1 cm* 0.8 cm and 2 cm* 0.5 cm* 0.4 cm. black papules were present over all three masses. An ulcer was seen over the largest mass. On histopathology, two of the lesions showed features similar to that of the initial biopsy. The largest lesion, however, showed two distinctive morphological features. Epidermis showed parakeratosis. Papillary dermis showed numerous ectatic and congested thin walled capillary channels in the papillary dermis close to the epithelium. Occasional fibrin thrombi were noted in these capillaries. This superficial part showed the features of solitary angiokeratoma (Figure 1 & 2). Deeper dermis showed a vague lobule composed of coiled eccrine glands closely intermingled with thin walled capillary channels, adipose tissue and pilosebaceous structures. At places, the superficial and deeper lesions appeared as a continuum. The deeper lesion was similar to the histopathological features of the two smaller lesions i.e. EAH (Figure 1 & 3). Since, these two morphological entities were seen in the same clinical lesion, a final diagnosis of EAKH was made.
DISCUSSION:

EAKH was recognized as a new entity by Kanitakis et al in 2006\cite{1}. They reported a lesion that met the clinicopathological criteria of EAH and surface of which showed features of angiokeratoma in an adult male patient and suggested the new nomenclature.

Clinically, EAH can presents as one or several nodules or a solitary large plaque. The lesions are generally present at birth and commonly involves an extremity\cite{2}. Hyperhidrosis and/or pain may be apparent. Pain is thought to occur due to involvement of nerve fibers, and hyperhidrosis because of the stimulation of the eccrine components, caused by the elevated local temperature within the angioma\cite{2,5}. Itching and hypertricosis have also been reported\cite{6}. On skin biopsy, the lesion is seen lying in the deep dermis and contain increased numbers of eccrine structures and numerous capillary channels surrounding or intermingled with the eccrine structures. Fatty tissue and pilar structures may also be seen intermingled with eccrine and vascular component\cite{2,7}. Intermingled abundant mucin has also been reported\cite{8}.

On the other hand, Angiokeratoma presents as tiny red papule on the skin surface. It is subcategorised on the basis of site of distribution in the body as Angiokeratoma corporis diffusum (trunk), Angiokeratoma of Mibelli (dorsum of fingers or toes), Angiokeratoma of Fordyce (scrotum), Solitary or multiple angiokeratomas (lower extremities). The first two subtypes are seen in childhood while the latter two commonly present in adults. Old and thrombosed lesions may appear blue-black and mimic malignant melanoma. A case of angiokeratoma on oral mucosa has also been reported\cite{9}. Histologically, all four types have same features consisting of numerous, dilated, thin-walled, congested capillaries mainly in the papillary dermis underlying an epidermis that shows variable degrees of acanthosis with elongation of the rete ridges and hyperkeratosis\cite{3}.

In the present case, out of the three lesions two showed classical features of EAH. However, the third lesion had features of both EAH and angiokeratoma, hence the diagnosis of EAKH. The vessels showed positivity for CD 31 and the epithelial cells of eccrine glands were positive for Cytokeratin on IHC. So far only two cases of EAKH have been reported including the first case which recognized this entity\cite{1,4}. Both these cases were seen in adults, one male and one female, with presentation since childhood. Both cases showed a superficial component of angiokeratoma and a deeper component of EAH. In a case series of 15 cases of EAH, Lin et al described one case associated
with features of angiokeratoma in a 32 years old male patient. However, they did not label the lesion as EAKH\cite{6}. In contrast, in our case the patient is a child and the lesion was present since birth. Also, the vessels in the EAH component of the present case are thin walled while in the case presented by Raghavan et al, they were thick walled.

Clinically, this lesion may mimic other congenital vascular lesions. Since, EAKH appears to progress with time, it is important to distinguish it from spontaneous regressing vascular lesions. Surgery is the treatment of choice for disfiguring or symptomatic cases.

**CONCLUSION:**

The case presented herein represents first documented congenital case of EAKH. EAH itself is an uncommon condition. EAKH is still rarer. Also, there is inadequate awareness due to its being a new entity. It needs to be distinguished from other common congenital vascular lesions. The congenital and childhood presentation of EAKH appears to be part of the same spectrum. However, the clinical and prognostic implications can be determined only after studying a larger number of similar cases.

**REFERENCES:**