Eccrine Angiomatous Hamartoma: A Rare Entity

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ABSTRACT

Eccrine angiomatous hamartoma (EAH) is an exceedingly rare benign tumor-like lesion, mostly develops before adulthood as single slow growing lesion on extremities, may produce pain and marked sweating. Presentations are manifold and the naevi are not always present from birth. The histological features include proliferation of eccrine sweat glands and angiomatous capillary channels. We report a 13 year old male who had a single lesion on his right thigh for last 4 years. Physical examination revealed a slightly elevated, 8x6 cm blackish-brown indurated, tender plaque on the posterolateral aspect of right thigh. Sweating in the lesion was evoked by physical work or emotional stress. Histopathological examination of the lesion showed increased numbers of eccrine glands, ducts and neural tissue as well as dilated vascular channels in the superficial and deep dermis. These findings are consistent with EAH. We report this case due to rarity of its presentation.

Keywords: Eccrine angiomatous hamartoma, Naevi

Introduction -

Eccrine angiomatous hamartoma (EAH) is a benign enlargement of eccrine components, accompanied by abundance of vascular channels. Increased proliferation of pilar structures, adipose tissue, and epidermis may be present. The anomaly is usually asymptomatic, but pain, hypertrichosis, and hyperhidrosis have been reported in a few patients (as in our case). Recurrence is rare. Differentiation from other angiomatous growths is necessary.

EAH also known as sudariparous angioma when there is a prevalence of angiomatous elements and eccrine elements are dilated, but not hyperplastic. However some authors consider these to be separate entities.

Case Report -

A 13 year old male presented with blackish brown plaque noted on his right posterior aspect of thigh for the last 4 years. His plaque was gradually increasing in size and now involved lateral aspect of thigh for last 2 year (Fig. 1). His lesion has grown proportionately with the patient. He was embarrassed noticing excessive sweating over elevated lesion, more so in summer. He also had pain in the lesion. Past medical history was insignificant. There was neither suggestive family history nor any history of trauma to the part. Physical examination revealed a slightly elevated, 8x6 cm blackish-brown indurated, tender plaque on the posterolateral aspect of right thigh. Sweating in the lesion was evoked by physical work or emotional stress. Histopathological examination of the lesion showed increased numbers of eccrine glands, ducts and neural tissue as well as dilated vascular channels in the superficial and deep dermis. These findings are consistent with EAH. We report this case due to rarity of its presentation.

X ray of both the hip joints and thigh was normal. MRI was suggestive of right Gluteus muscle atrophy (Fig. 2). A 4 mm punch biopsy was performed and histopathology revealed a thickened epidermis and an increased number of small and medium-sized bloodvessels with thin muscular walls of uneven thickness (Fig. 3). An increased number of eccrine glands with large coils composed of an increased number of tubules and ducts with a normal proportion of cells were noted in the dermis (Fig. 4). Repeated cultures of biopsied samples showed no...
growth of fungi, mycobacteria or bacteria, suggesting that inflammation was not caused by infection. The outcome of the clinical and histopathological finding pointed conclusively to eccrine angiomatous hamartoma. The patient was referred to plastic surgery department for surgical management.

**Discussion**

EAH was first described by Lotzbeck in 1859 as an angiomatous appearing lesion on the cheek of a child. The term EAH was coined by Hyman and coworkers in 1968.

EAH occurs with an equal incidence in both sexes, with approximately one third of the cases manifesting at birth or in early childhood. There are few reports of adult-onset cases up to age 71. The etiology of EAH has not been clearly defined. According to Zeller and Goldman it might becaused by the abnormal induction of heterotypic dependency during organogenesis. Clinically, EAH presents as an angiomatous lesion, usually solitary, although cases with multiple lesions have been described. Clinical manifestations may vary from nodules to plaques of erythematous bluish or brownish color or verrucous lesions mostly located on the extremities (80%), however, other locations like trunk, neck, face, and vulva have been reported. EAH, generally asymptomatic, may present with focal hyperhidrosis and pain that is spontaneous or follows local pressure. The pain occurs due to involvement of nerve fibers and hyperhidrosis is because of the stimulation of the eccrine components, caused by the elevated local temperature within the angioma. Generally, they enlarge very gradually; more rapid growth has been described during pregnancy and adolescence suggesting underlying hormonal influence.

On histopathology, EAH is characterized by a dermal proliferation of well-differentiated eccrine secretory and ductal elements closely associated with thin-walled angiomatous channels. In addition, unusual histopathological variants have been reported and include the infiltration of adipose tissue, increased dermal mucin, and the presence of apocrine glands or pilar structures. Where the angiomatous element predominates, the term sudoriparous angioma has been applied. The nevus sudoriparous type of eccrine nevus may represent the non-angiomatous end of the same spectrum of hamartomas.

Clinically, this condition must be differentiated from other neonatal angiomatoses. Sometimes the clinical findings are nonspecific, whereas histological examination may exclude the other conditions. The differential diagnosis of EAH may include smooth muscle hamartoma, an eccrine nevus, tufted angioma, macular telangiectatic mastocytosis, nevus flammeus, Kimura’s disease, pyogenic granuloma and glomus tumor.

Kimura's disease is a classical triad of painless subcutaneous masses in the head and neck region, blood and tissue eosinophilia and markedly elevated IgE.

Pyogenic granuloma occurs in children and young adults with usual presentation being solitary, well circumscribed, dome shaped, 1-10 mm, sessile or pedunculated, bright or dusky red, smooth, firm nodule. Most lesion are asymptomatic. Histopathology reveals a lobular pattern separated by fibrous septa. Each lobule consists of capillaries and venules lined by plump endothelial cells and embedded in an edematous gelatinous stroma.

While definitive diagnosis is based upon histology, MRI and ultrasound are useful to delineate the extent of soft tissue and vascular masses. Immunohistochemical study (not performed in this case) can demonstrate that antigens frequently found in eccrine glands, such as the carcinoembryonic antigen and protein S-100, are reduced qualitatively in the eccrine glands of EAH.

The natural history of EAH is benign and typically slow-growing hence aggressive treatment is generally unwarranted. Simple excision is usually curative and is reserved for painful and cosmetic disfiguring lesions. Deep excision with full-thickness grafting, or amputation of a finger or toe may be required for symptom control in those with
larger lesions on acral parts. Pulsed-dye laser and Nd : YAG laser treatment have been performed without much success. Recently botulinum toxin and intraleisionalsclerosants have been successfully used in the treatment of this condition. It is important to recognize this condition because it is a benign lesion for which aggressive treatment is not indicated. This case is being reported as very few cases have been reported from India and to the best of our knowledge this is the first case associated with muscle atrophy.

Conflicts of interest: None reported by Authors

References:


