Eccrine Angiomatous Hamartoma: A Rare Presentation in Late Adulthood

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Abstract:

Eccrine angiomatous hamartoma is a tumor characterized by benign malformation of eccrine and vascular elements, occurring generally at birth or during early infancy and childhood. The rarity with which it is encountered in adults and the elderly makes it a diagnostic challenge in such age brackets. We report an interesting case of eccrine angiomatous hamartoma presenting in late adulthood confirmed by histopathology.

Keywords: Cisplatin, concurrent chemotherapy, daily, weekly, three weekly, carcinoma cervix

Introduction:

Eccrine angiomatous hamartoma (EAH) is a rare, benign lesion histologically characterized by hyperplastic eccrine and vascular structures which may also be associated with proliferation of other dermal elements like hair follicles and adipose tissues. EAH was first described by Lotzbeck in 1859 and the term was coined by Hyman et al in 1968^[1,2]. It arises at birth or appears in early childhood, with only a handful of reports of adult onset lesions documented in literature.

Case Report:

A 57-year-old male patient presented with complaints of swelling on the second toe of right foot for duration of one year. It was gradually progressive in size. There was no history of local trauma, hypertrichosis, hyperhidrosis, any

systemic illnesses or similar swelling in any other part of the body. Patient denied history of similar lesions in his family. Local examination showed an ill-defined swelling measuring approximately 4cm x 2.5cm which was soft and non-tender. The overlying skin was normal. His routine blood parameters were within normal limits. An ultrasonogram of the swelling revealed a cystic lesion with few internal septations showing no significant flow on color doppler. CEMR right foot revealed a lobulated cystic lesion on superior aspect of second digit (also extending along medial margin) overlying proximal and middle phalanges, being hypointense on T1W and hyperintense on T2W images with slight peripheral enhancement on CEMR and rim hypointense on T2W images which was thin and regular. A clinical and

radiological diagnosis of giant cell tumor of tendon sheath was rendered. Following which an excision biopsy was performed and submitted for histopathology.

We received a grey white soft tissue piece measuring 2cm x 1cm x 0.5cm. Microsections

processed and examined showed an intricate mixture of fibrocollagenous, fibroadipose tissue with proliferating thick and thin-walled blood vessels admixed with proliferating eccrine glands and ducts (figure1-3). Based on these findings, a diagnosis of EAH was made.

Figure 1: Proliferating eccrine glands and abnormally dilated vascular channels adjacent to fatty lobules. (H&E, X20)

Figure 2: Increased number of eccrine glands associated with clusters of blood vessels. (H&E, X20)

Figure 3: Proliferating variably sized angiomatous vascular structures and eccrine glands in a fibrocollagenous stroma. (H&E, X20)

Discussion:

Eccrine Angiomatous Hamartoma (EAH) is a rare benign cutaneous malformation, comprising of proliferating eccrine secretory coils and ducts associated with capillary angiomatous channels and other minor structures like fatty lobules and hair follicles. EAH was at first described as sudoriparous angioma by Lozbeck in 1859.^[1] Later on, sudoriparous angioma was categorized by some authors as a distinctive condition.^[3] Eccrine nevus and sudoriparous angioma may demonstrate certain histologic overlap, although in the former, hyperplastic eccrine glands are not associated with a proliferation of capillaries and most eccrine nevus demonstrate hyperhidrosis; whereas in sudoriparous angioma, the angiomatous component predominates over the dilated but not hyperplastic eccrine elements.^[4]

EAH characteristically appears at birth or during early childhood, while in our case it presented at a much later age. EAH commonly manifests as patches, plaques, or nodules. It may present as a solitary or as multiple lesions showing predilection for distal extremities where there is increased number of eccrine glands. Sulica et al proposed that the lesion may be associated with tenderness due to compression of small nerve branches by the hamartomatous structures, rather than actual proliferation of neural elements in the lesion.^[5]

Although the definitive etiology underlying EAH remains undetermined, it has been suggested by Zeller and Goldman that the growth arises due to an aberration between the differentiating epithelium and the underlying mesenchyme resulting in the abnormal proliferation of adnexal and vascular structures.^[6] In children, EAH typically progresses with patient's growth, which is characteristic of a hamartoma, yet some incidences of rapid increase in size during puberty and pregnancy suggests a role of hormonal stimulation.^[7] The reported cases of adult onset EAH have shown correlation with recurrent trauma and irritation, however our case does not correlate with the same.^[6, 8]

Microscopic evaluation is essential for definite diagnosis of EAH because the clinical features are

nonspecific and variable. The histologic criteria for the diagnosis of EAH proposed by Pelle et al includes: (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.^[9]

The usual differential diagnosis of EAH includes tufted angioma, vascular malformations, macular telangiectatic mastocytosis, nevus flammeus, glomus tumor, giant cell tumor, smooth muscle hamartoma, congenital hamartoma of the eccrine sweat gland, eccrine nevus, eccrine poroma and blue rubber bleb nevus, all of which can be differentiated by their respective histopathology features.^[10]

In light of the overlapping features that these other lesions share with EAH and their variable clinical presentations, distinction can prove to be a difficult task even on biopsy and for that reason, some cases should be approached descriptively with the need for clinicopathologic correlation.

Due to its benign nature and slow progression, aggressive treatment is unnecessary. In cases of disabling symptoms or for cosmetic reasons, surgical excision remains the mainstay of treatment.^[11]

Conclusion:

Eccrine angiomatous hamartoma although rare in adulthood should be included in the differential diagnosis of cutaneous nodules. Awareness of the variable clinical presentations with wide age spectrum of this condition can aid in diagnosing this entity in which vascular and eccrine anomaly serve as primary pathogenetic events.

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