Case Report

Eccrine Angiokeratomatous Hamartoma: Report of A Case with Brief Review

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ABSTRACT

Eccrine angiokeratomatous hamartoma is a newly recognised vascular lesion of the skin, with the first case being reported only in 2006. It has a distinctive histology with features derived from two well established vascular lesions affecting the skin: solitary angiokeratoma and eccrine angiomatous hamartoma.

A 26 year old woman presented with a long standing painless warty lesion on her right ankle. Histological examination of the excised lesion showed a combination of two vascular lesions: superficial angiokeratoma and a deeper eccrine angiomatous hamartoma. Such a histological picture is consistent with the diagnosis of recently described eccrine angiokeratomatous hamartoma. Our case represents only second such case to be reported. Immunohistochemical analysis with CD34, smooth muscle actin and cytokeratin was done.

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Introduction

Eccrine angiokeratomatous hamartoma (EAKH) is a newly recognised vascular lesion of the skin, with the first case being reported only in 2006[1]. It has a distinctive histology with features derived from two well established vascular lesions affecting the skin: solitary angiokeratoma (SAK) and eccrine angiomatous hamartoma (EAH). Solitary or multiple angiokeratoma is a very common condition that presents as a single or many papular lesions on the extremities of young adults [2]. EAH is relatively rare lesion that is usually present at birth or manifests early in the childhood as multiple nodules or as a single large plaque on the extremities [3]. Since its recognition, no other case of eccrine angiokeratomatous hamartoma has been documented until now. We are reporting second such case.

Case Report

A 26 year-old woman presented with a painless, flesh-coloured warty lesion over the lateral malleolus of her right ankle. She had no history of trauma or surgery in the recent past and nor did she suffer from any other major illness. The patient first noticed this lesion when she was 4 years old, when it apparently started as a tiny painless nodule. Since then, it had grown very gradually to its present size of 3 cm. On several occasions in the past, it ulcerated following trivial trauma and was accompanied by pain, tenderness and crusting. No one else among her immediate family had similar lesion.

The lesion was surgically excised and was submitted for hisopathological examination. The excised specimen consisted of a thick elliptical piece of skin with subcutaneous tissue. It measured 3.5x2x2 cm. The skin was verrucous. On bisection, the cut surface was fairly homogeneous tan-white, but no clearly demarcated lesion was seen.

Entire specimen was processed for hisopathological examination. Besides the routine haematoxylin and eosin (H&E) staining, the sections were also subjected to immunohistochemistry with antibodies against CD 34, cytokeratin, desmin and smooth muscle actin-a (SMA-α) [Novocastra (Leica) Mouse monoclonal antibodies].

On microscopic examination, the epidermis showed verrucous hyperplasia. But the histology was dominated by a poorly delineated vascular lesion with two distinctive morphological areas: The superficial part, which involved papillary and superficial reticular dermis, was composed of thin walled, partly ectatic capillary vessels, which at places were partly enclosed within an epidermal collarette. Some of the larger vessels were occluded by fresh intraluminal fibrin thrombi. This portion had the features of solitary angiokeratoma

The deeper portion had a distinctive lobular configuration and affected the mid and deep reticular dermis. It was composed of fairly uniform thick walled capillaries intricately admixed with sweat ducts and glands. Some of the lobules contained adipose tissue with myxoid interstitium. Some of the deeper vessels showed intraluminal fibrin thrombi. This part had the histological features of eccrine angiomatous hamartoma.

Immunohistochemically, the proliferating blood vessels in both areas were positive for CD 34 (figure 3 A & B) and smooth muscle actin-α (figure 4 C & D). The latter was positive either as a continuous or discontinuous single layer within the walls of vessels in the angiokeratomatous area. In the deeper eccrine angiomatous hamartomatous area, it was thick, multi-layered and more abundant. Desmin was negative in all the proliferating vessels. Cytokeratin delineated the eccrine glands and ducts in the deeper part (figure 5).

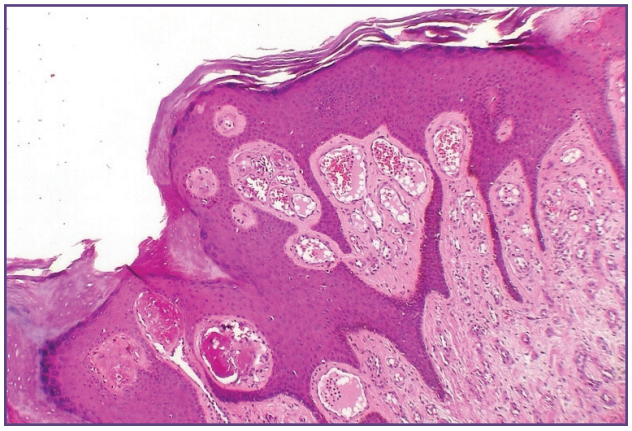

Fig. 1: From the superficial part of the lesion showing Angiokeratoma-like area (Haematoxylin and Eosin, 100X)

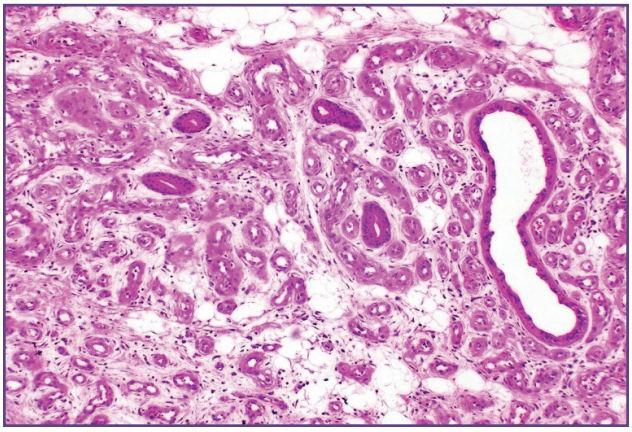

Fig. 2: From the deeper part of the lesion showing Eccrine Angiomatous Hamartoma-like area (Hearmatoxylin and Eosing, 100X)

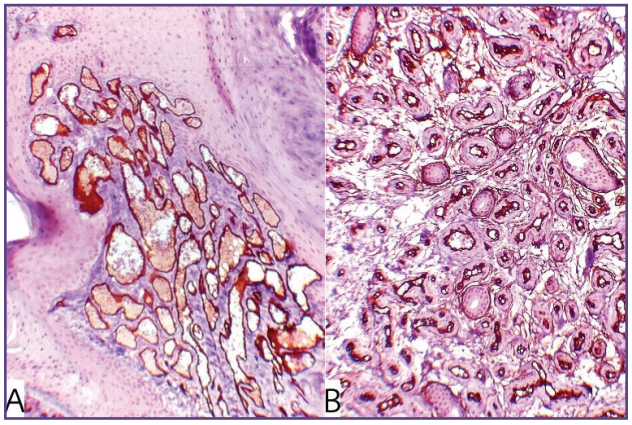

Fig. 3: Immunohistochemistry for CD34. Blood Vessels in superficial (A) and deeper (B) portions of the lesion are strongly positive

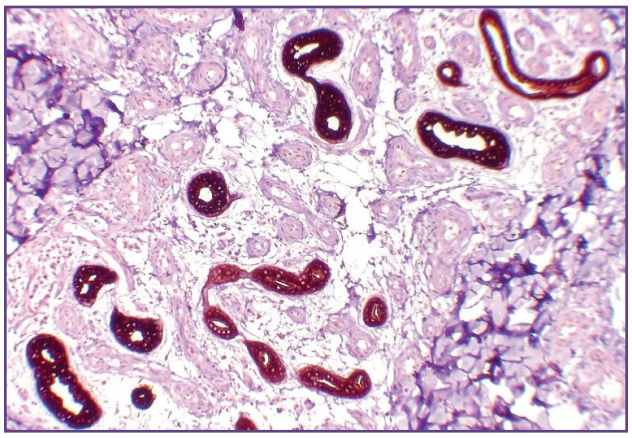

Fig. 5: Immunohistochemistry for cytokeratin. Sweat glands and ducts in eccrine angiomatous hamartoma-like area are positive.

Discussion

EAKH was recognised as a distinct new entity in 2006 by Kanitakis et al [1] when they discovered that a case clinically diagnosed as EAH had histological features of both SAK and EAH. They proposed the new name as well. In their case too, the clinical presentation was similar to the present case and the lesion was seen on the right malleolus. It was small but tender.

In the present case, the patient noticed the lesion as a small nodule when she was 4 years old. Its progression was very slow and was punctuated by several episodes of painful ulcerations. At the time of presentation, the lesion measured only about 3 cm and was painless. Histologically, as we have described, it exhibited features of both angiokeratoma and eccrine angiomatous hamartoma. The blood vessels in these two areas were morphologically different. The angiokeratomatous areas had thin, ectatic capillaries of

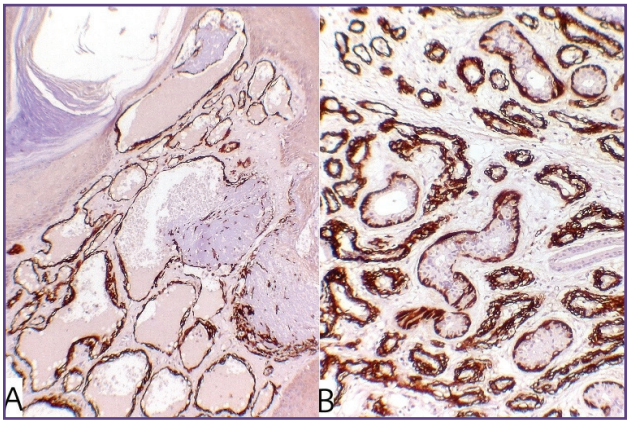

Fig. 4: Immunohistochemistry for SMA-alpha. Blood vessels in superficial (A) and deeper (B) portions of the lesion are strongly positive

varying sizes (figure 1). But the deeper EAH-like areas had fairly uniform small thick walled vessels resembling arterioles (figure 2) unlike what was reported by Kanitakis et al [1]. Immunohistochemically, the vessels in both areas expressed CD 34 and SMA-α but the intensity of staining for SMA-α was more in the EAH-like area (figure 3 & 4). As SMA-α is expressed by not only smooth muscle cells but also by the pre-and post-capillary pericytes and myofibroblasts [4], immunohistochemistry for desmin was done, as it is expressed only by muscle cells and can be used to distinguish between pericytes and smooth muscle cells. All the proliferating vessels in our case were negative for desmin. So, even though the vessels in deeper portion appeared thick walled, they did not have any smooth muscle in their walls and the thickness observed is probably due to proliferation of pre- and post- capillary pericytes. In both areas, some of the blood vessels were occluded by fresh fibrin thrombi, a feature frequently observed in SAK [2].

The differential diagnosis for this would be EAH and SAK. Eccrine angiomatous hamartoma (EAH) is a relatively rare condition that usually manifests at birth or early in childhood as a papule or a nodule or a plaque on distal portions of the extremities [5] [6] [7] [8]. But they have been reported on other sites including head and neck region and trunk [9] [10]. It has also been reported in adults [11]. It commonly presents as a solitary lesion, but occurrence of multiple EAH has been documented [12] [13]. The lesion is usually asymptomatic but may be associated with pain and hyperhidrosis [14]. Histologically the lesion affects mainly mid and deep dermis and is composed of lobules of proliferating capillaries intricately admixed with sweat glands and ducts, fat and myxoid tissue. Immunohistochemically, the sweat glands have been shown to be positive for S 100, EMA and cytokeratin; and the vascular component for CD 31 and CD 34.

Solitary or multiple angiokeratoma (SAK) is a common condition that tends to affect the lower extremities of young adults. However, it has been reported from other sites as well including oral cavity [15]. It usually presents as solitary or multiple papular lesions. The histology is characterised by the presence of thin ectatic capillary vessels in papillary dermis with overlying epidermis showing verrucous hyperplasia. The rete ridges at places may form epidermal collarette. Some of the vessels may show fresh thrombi and the vessels are usually positive for CD 31 and CD 34.

Since its recognition in 2006, no other case of EAKH has so far been documented. As our case meets the morphological diagnostic criteria of EAKH, it represents only the second such case to be recognised. Paucity of documentation reflects either the rarity of this condition or lack of awareness. Whether it is distinctive entity or a variant of EAH or SAK can only be established if a few more cases are reported.

Conclusion

We are presenting only the second documented case of eccrine angiokeratomatous hamartoma having morphological features of both angiokeratoma and eccrine angiomatous hamartoma. Whether it is a distinctive entity or a variant of those two lesions remains to be established.

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Competing Interests

None declared

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