

CASE REPORT

Eccrine Angiomatous Hamartoma Mimicking a Traumatic Hemorrhage

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Eccrine angiomatous hamartoma (EAH) is a rare benign disease that is characterized by an abnormal proliferation of eccrine glands and vascular elements. It is generally congenital, but it can appear before puberty. It usually presents as a single plaque or nodule, but multiple patch-like lesions are also possible. EAH is mostly asymptomatic, but it is sometimes associated with pain or hyperhidrosis. It generally does not require aggressive treatment, but the lesion can be excised due to pain, enlargement and cosmetic reasons. A 3-week-old Korean female presented with a hemorrhagic skin lesion on the right foot since birth. There was no specific birth history. The lesion first appeared on the third toe of the right foot and quickly spread to almost half of the right foot. Histopathology examination revealed acanthosis in the epidermis and a proliferation of eccrine ducts, glands and capillaries. The eccrine glands were immunohistochemically-positive for carcinoembryonic antigen. (**Ann Dermatol 23(S1) S84 ~S87, 2011**)

-Keyword-

Eccrine glands, Hamartoma

INTRODUCTION

Eccrine angiomatous hamartoma (EAH) is a rare disease that is histologically characterized by increased numbers of eccrine structures and numerous capillary channels¹. It commonly appears as a flesh-colored, red, violaceous, blue, or yellow-colored solitary nodule or plaque on the distal extremities, sometimes presents as multiple papules, and rarely presents as maculopatches on the trunk, buttocks, and neck. It may be congenital or it can arise before puberty, but the lesion rarely presents in adulthood. EAH is mostly asymptomatic, but pain, hypertrichosis, and local hyperhidrosis are commonly reported features. Enlargement typically occurs and this is associated with growth of the patient. Although it is a benign hamartoma, it is often excised because of pain or enlargement and for cosmetic reasons².

We report here on a case of a congenital EAH that appeared as fast-growing hemorrhage-like maculopatches.

CASE REPORT

A 3-week-old Korean girl presented with well-demarcated



Fig. 1. Well-demarcated hemorrhage-like maculopatches evident on the right foot.

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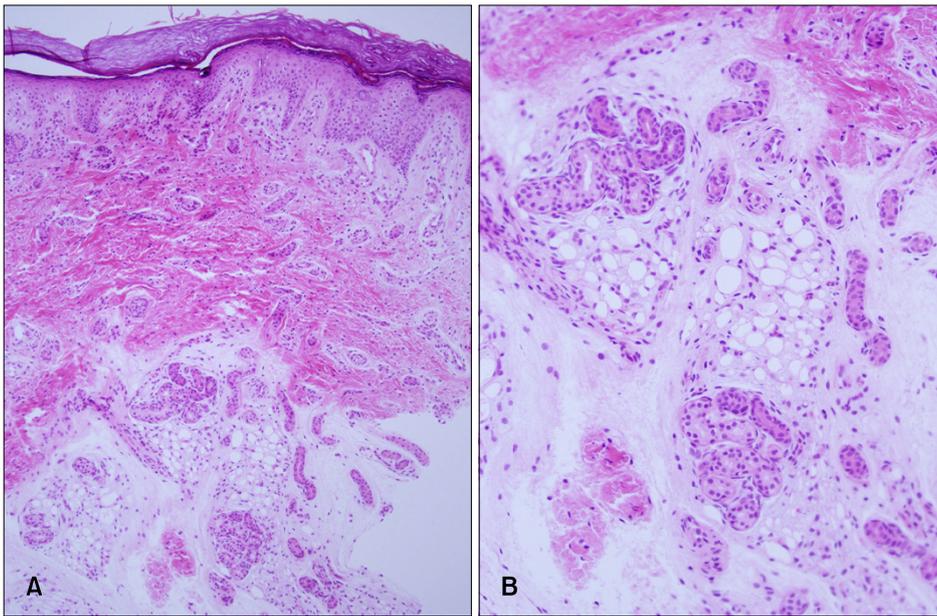


Fig. 2. Results from a skin biopsy. (A) The biopsy specimen shows irregular acanthosis with hyperplasia of the eccrine glands and capillaries in the mid-upper dermis (H&E, $\times 100$). (B) Hyperplasia of the eccrine glands and capillaries associated with fatty tissue (H&E, $\times 200$).

hemorrhage-like maculopatches on the right foot since birth, and this lesion had appeared as a small hemorrhagic macule on the third toe of the right foot and then quickly spread over the ensuing 1 week. There was no specific birth history or trauma history. On examination, the lesions were revealed to be erythematous petechiae-like maculopatches that covered almost half of the right sole dorsum and toes. The condition was seemingly painless with palpation and it did not appear to be hypertrichotic (Fig. 1). The mother of the patient reported that there was no hyperhidrosis. A 4-mm punch biopsy was done. In the subsequent histopathology, slight acanthosis was found in the epidermis. In the dermis, there were numerous hyperplastic eccrine glands and capillaries nearby, but no pilar, lipomatous, mucinous, or lymphatic structural hyperplasia (Fig. 2). The eccrine glands were positive for carcinoembryonic antigen (CEA) on immunohistochemistry examination (Fig. 3). Based on the histopathological features, the lesion was diagnosed as EAH. The treatment options presented to the parents included observation or surgical excision. The decision was for observation.

DISCUSSION

EAH is a rare benign tumor of the skin, and it is defined histopathologically as eccrine hyperplasia with a proliferation of vascular elements and, in some cases, the presence of other dermal elements such as mucin or lipid. The first of these lesions was documented in 1859 in the description of an angiomatous tumor located on the cheek of a child². The term EAH was coined in 1968³. Other

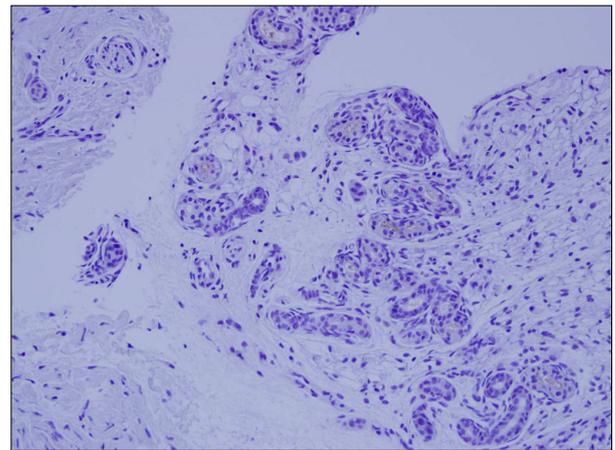


Fig. 3. Positive CEA immunostaining of the eccrine ducts and secretory lobules ($\times 200$).

terms such as sudoriparous angioma were used in prior reports³. A pathophysiologic theory to explain EAH involves a biochemical fault in the interactions between the differentiating epithelium and subjacent mesenchyme that gives rise to an abnormal proliferation of adnexal and vascular structures⁴.

A study that reviewed 37 EAH cases revealed the generally congenital nature of EAH (16 of 35 cases, 45.7%; two cases were not mentioned) and its prevalent appearance at a prepubertal age (21 of 36 cases, 58%; one case was not mentioned)². Although it rarely occurs in adulthood, there have been some reported cases of late-onset EAH⁵. Results from the same study indicate that a single nodule (32/37, 86.5%) or plaque (28/37, 75.6%)

Table 1. Reported cases of eccrine angiomatous hamartoma in Korean literatures

Author	Age/sex	Onset	Sweat	Pain	Location	Structure	No. of lesion
Present case	1 mo/F	Birth	No	No	Right foot	Patch	Single
Ahn et al. ⁶ (1981)	13 mo/M	Birth	No	No	Right knee	Nodule	Single
Kim et al. ⁷ (1982)	18/F	Birth	Yes	Yes	Right leg	Nodule	Multiple
Kim et al. ⁸ (1982)	16/F	6 yr	Yes	Yes	Left buttock	Nodule	Multiple
Choi et al. ⁹ (1982)	5/F	Birth	Yes	No	Left thigh	Patch	Single
	9/F	Birth	Yes	Yes	Right leg	Nodule	Multiple
	4/M	Birth	Yes	Yes	Right knee	Nodule	Single
Lee et al. ¹⁰ (1988)	5/F	3 mo	Yes	Yes	Right knee	Nodule	Single
Ryu et al. ¹¹ (2002)	2/M	Birth	Yes	No	Right knee	Nodule	Single
Choi and Chung ¹² (2003)	10 mo/M	10 mo	No	No	Right knee	Nodule	Single
	34/M	29 yr	No	No	Left shoulder	Nodule	Single
Yoon et al. ¹³ (2006)	16/F	X	Yes	Yes	Left forearm	Nodule	Single
Kwon et al. ¹⁴ (2008)	6/F	Birth	No	No	Right knee	Nodule	Single
Kim et al. ¹⁵ (1984)	10 mo/F	1 mo	Yes	No	Right thigh	Plaque	Single
Kwon et al. ¹⁶ (1998)	50/M	Birth	Yes	No	Left chest	Patch	Single

X: not mentioned.

is the dominant lesion, but multiple nodules (5/37, 13.5%) or papulopatches (12/37, 32.4%) are also possible, and that EAH is mostly located in the extremities (30/37, 81.1%). Considering the reported clinical symptoms, EAH has various features. Results from the aforementioned study indicated an asymptomatic nature (12/33, 36.3%), but it can also present with pain (14/33, 42.4%), hyperhidrosis (12/35, 34.3%) or hypertrichosis (10/37, 27.0%). There is no gender preference. The same clinical features were evident in five congenital cases^{2,4}. The clinical differential diagnoses of EAH include vascular malformations, tufted angioma, smooth muscle hamartoma, glomus tumor, blue rubber bleb nevus, and macular telangiectatic mastocytosis².

In addition to the present data, we also reviewed the results from 14 Korean cases⁶⁻¹⁶. A summary of the results is presented in Table 1; the clinical features were almost the same clinical features as those described in an English review article². The present and previous Korean patient data were obtained from slightly more females (n=9) than males (n=6). The lesions were mostly congenital (9/14, 64.2%), and lesions could be asymptomatic (5/15, 33.3%) or painful (6/15, 40.0%), and excessive sweating was evident (10/15, 66.7%). A single lesion was typically present (12/15, 80.0%) and almost always as a nodule (11/15, 73.3%). But, three cases that presented as a patch, including our case. Lesions appeared on the extremities in most cases (12/15, 80.0%). Clinically, our case was quite consistent with most of the reported cases, except for the appearance as a skin patch instead of as nodules or plaques. Although two previous cases displayed a lesion patch, similar to the present case, they did not show

traumatic hemorrhage-like maculopatches as in our case. Rather, the lesions of the two previous cases appeared like a nevus flammeus eruption^{8,16}. The present case could represent a truly new type of EAH lesion. Alternatively, it may have not fully developed to become a nodule or plaque because the patient was too young. The differential diagnoses of hemorrhage-like maculopatches in infants include hematologic disorder, such as leukemia and Langerhans cell histiocytosis, hemangioma, and true traumatic hemorrhage.

Histologically, EAH displays variable epidermal findings such as normal or irregular acanthosis, papillomatosis and/or hyperkeratosis, hyperplasia of normal or dilated eccrine glands in close association with capillary angiomatous foci, and the variable presence of pilar, lipomatous, mucinous, and lymphatic structures^{2,4}. There was mild acanthosis in the epidermis and hyperplasia of the nearby normal eccrine glands and capillaries in our case. These findings match with those of most of the previous cases.

Immunohistochemical analyses of EAH has demonstrated that the secretory portions of the eccrine glands are positive for S100, CEA, epithelial membrane antigen, and Cam5.2, and that the ductal components stain positively for CEA and cytokeratin 1, and are weakly positive for epithelial membrane antigen². CEA staining done presently was also positive on the ductal and secretory portions of the eccrine glands.

In summary, we report here on a case of congenital EAH that had the appearance of a hemorrhagic lesion. To the best of our knowledge, this is the first case of EAH showing such unique clinical features.

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