Acta Dermatovenerol Croat 2024;32(2):102-104 CASE REPORT

A Pediatric Case of Multiple Bilateral Symmetric Eccrine Angiomatous Hamartoma

Ilke Beyitler, Fikret Dirilenoglu

Near East University, Faculty of Medicine, Nicosia, Cyprus

Corresponding author:
Prof. Ilke Beyitler
Near East University, Faculty of Medicine
Near East Boulevard, Nicosia, Cyprus
ilkebeyit82@yahoo.com

Received: September 26, 2023
Accepted: February 28, 2024

ABSTRACT
Eccrine angiomatous hamartoma (EAH) is a rare benign skin neoplasm characterized by an increased size and number of eccrine glands or ducts, along with proliferation of vascular structures in the dermis. This case is unique in its presentation of bilateral symmetrical nodules on both hands and the development of new nodules during puberty. It highlights the need for further research and understanding of this rare condition and its potential progression over time.

KEY WORDS: eccrine angiomatous hamartoma, multiple, symmetric

INTRODUCTION

Eccrine angiomatous hamartoma (EAH) is a rare benign skin neoplasm that was first described in 1859, with around 100 reported cases to date. The lesions are characterized by an increased size and number of eccrine glands or ducts, along with proliferation of vascular structures in the dermis. Lesions can be congenital or appear in infancy, childhood, or adulthood, with no gender predominance. The nodules or papules can be red, blue, yellow, violaceous, brown, skin-colored, or have variable sizes ranging from 1-25 cm (1-3). Solitary lesions are more common than multiple manifestations. The most common localization sites are the extremities, while the head, neck, and trunk are rarely affected. Although EAH cases are generally asymptomatic, pain, hyperhidrosis, walking difficulty, and hypertrichosis have also been observed (4,5).

In this report, we describe a rare presentation of bilateral multiple symmetric EAH beginning on four distal interphalangeal joints (DIP), one proximal interphalangeal joint (PIP), and with new lesions appearing on three other proximal interphalangeal joints (PIPs) of both hands during puberty. This case is unique in its presentation of bilateral symmetrical nodules on both hands and the development of new nodules during puberty.

CASE REPORT

An 11-year-old female patient presented with complaints of painful swellings on the fingers of both hands lasting for six months, recurrent bilateral knee pain, and abdominal colic-style pain lasting nine months. She had a history of appendectomy, and her family history revealed that her grandmother had nodules on her hands. Physical examination revealed bilateral, subcutaneous, dorsal, interphalangeal, slightly red-colored, soft, and painful nodules, measuring 0.5-1.0 cm in size, on the DIPs of the 2nd, 3rd, 4th, and 5th digits and the PIP of the 5th digit of both hands (Figure 1). Laboratory investigations, including complete blood count, erythrocyte sedimentation rate, C-reactive protein, liver, kidney, and thyroid function tests, 25(OH) vitamin D, routine urine analysis, immunoglobulin G, A, and M, antinuclear antibody, double-stranded DNA, tissue transglutaminase IgA, *MEFV* gene analysis, abdominal ultrasound, skeletal survey bone X-rays, and bilateral knee MRI were all normal. Histopathological examination of a

Acta Dermatovenerol Croat
2024;32(2):102-104

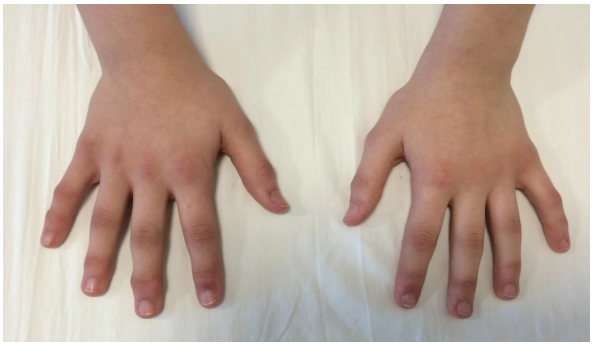

Figure 1. Nodules on the distal interphalangeal joints of the 2nd, 3rd, 4th, and 5th digits and the proximal interphalangeal joint of the 5th digit of both hands.

surgically excised nodule revealed proliferation of hamartomatous eccrine glands and blood vessels between and around the glands and ducts consistent with EAH (Figure 2). The patient's abdominal and knee pain subsided. She was followed up for 5 years, with the same nodules persisting without enlargement during puberty. However, new nodules have developed on the PIPs of the 2nd, 3rd, and 4th digits of both hands since she was 14 years old (Figure 3). The patient experienced pain while writing with her right hand and was recommended to use analgesics until excision of the other pain-causing lesions.

DISCUSSION

EAHs typically manifest in early childhood and are predominantly located on the extremities. The majority of cases present with a single lesion (6,7). The congenital form of EAH is thought to arise from malformations involving adnexal and mesenchymal elements, although the role of mechanical factors such as repetitive trauma in late-onset cases has been questioned. In our case, the patient's grandmother had a history of hand nodules, but her diagnosis is unknown. Familial presentation has been reported in one case involving an 11-year-old boy and his mother who both had multiple lesions. Although EAH is a benign condition, it was recommended that this genetic association be kept in mind (8,9).

Rare presentations of EAH have been documented, including a case where the lesion occurred at the site of recurrent excision of a squamous cell carcinoma in an 84-year-old patient, suggesting atypical proliferation on deformed eccrine and vascular arrangement (7). Another atypical onset of EAH occurred in a 14-year-old girl with antiphospholipid syndrome, who presented with a plantar violaceous plaque and foot pain. A biopsy showed proliferation of eccrine glands as well as capillaries with microthrombi. Altered endothelial integrity and vascular flow as a consequence of dermal malformations were examined (10). Arteriovenous malformations were also detected in biopsy specimens of a few EAH cases, expanding the histological findings of this entity (11). In addition, tumorigenic growth factor is present in the skin of patients with neurofibromatosis type-1, which plays a role in angiogenesis, neurogenesis, and cell migration, and was suggested as having a possible role in the vascular proliferation of EAH (9).

To date, only five cases of bilateral symmetric EAH have been reported. An adult male had bilateral lesions on the backs of both fingers on the hand, and a 14-year-old adolescent had bilateral papules on the backs of both thumbs (12). A 14-year-old girl had nodules on the extensor sides of both wrists that were excised due to pain (13). Another adolescent case had bilateral painless symmetric nodules on the volar sides of the wrists, without recurrence 23 months after excision (1). An adult case had bilateral dorsal lesions, initially on three PIPs, that over time expanded to involve all five PIPs bilaterally (14).

Spontaneous regression of EAH has been observed, particularly in infants, indicating that a watch-and-wait approach may be suitable for asymptomatic cases (15). In contrast, there have been instances where EAH lesions that were asymptomatic for years suddenly enlarged and became painful. One such case involved an adolescent girl who experienced these symptoms in a toe lesion approximately one year after menarche (16). This sudden change was

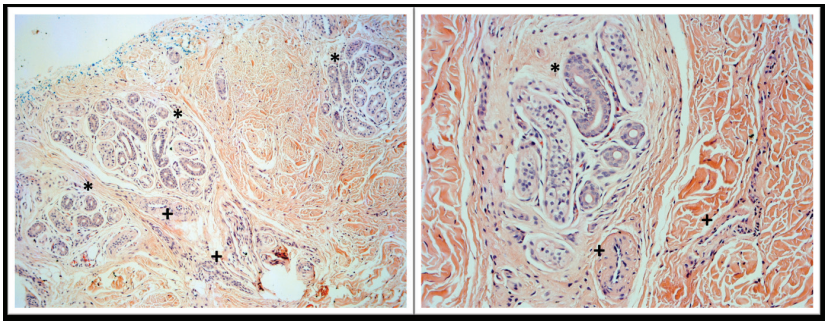

Figure 2. Histopathological examination revealed proliferation of hamartomatous eccrine glands (*) with blood vessels (+) between and around the glands and ducts (hematoxylin and eosin, ×100 (left) and ×400 (right)).

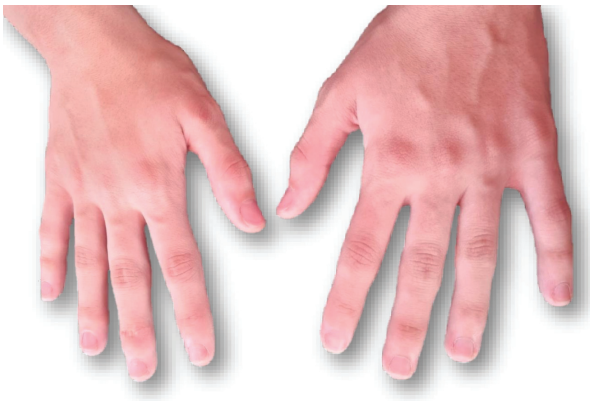

Figure 3. Nodules that developed later on the proximal interphalangeal joints of the 2nd, 3rd, and 4th digits of both hands.

attributed to hormonal stimulation during puberty, suggesting that hormonal changes can influence the growth and pain associated with EAH (8,15,16).

The current case is unique in that it presented with 16 bilateral symmetrical nodules on four DIPs and one PIP, with the subsequent development of bilateral symmetrical nodules on three other PIPs. Although enlargement of EAH lesions during the growth of children, puberty, and pregnancy has been rarely reported, an increase in the number of lesions over time is an even rarer presentation of this entity. It highlights the need for further research and understanding of this rare condition and its potential progression over time.

Ethical approval:

Informed consent was received from the family.

Source of funding:

The authors declare that no funding was received for this study.

Conflict of interest:

The authors declare that there is no conflict of interest.

References:

1. Clayton A, McCarthy EF, Puttgen KB, Deune EG. Bilateral eccrine angiomatous hamartoma with symmetric volar wrist nodules. JBJS. 2019 Jan-Mar;9:e20.
2. Finnigan CJ, Conejero C, Martin AH, Gomez JS, Morel LN. Congenital erythematous plaques and papules on the right arm. Pediatr Dermatol. 2015;Mar-Apr;32:285-6.
3. Nygaard U, Dalager S, Spaun E, Hedelund L. Large eccrine angiomatous hamartoma: a novel clinical presentation of disease. JDCR. 2015;3:58-61.
4. Hernot S, Wadhera R, Kaintura M, Bhukar S, Pillai DS. Angiomatous hamartoma – A rare presentation. J Clin Diagn Res. 2016 Sep;10:MD04-MD05.
5. Halder C, Naskar B, Biswas P, Mitra PK. Eccrine angiomatous hamartoma: Late onset facial presentation. Indian J Dermatol. 2014 Jul-Aug;59:403-5.
6. Sanusi T, Li Y, Sun L, Wang C, Zhou Y, Huang C. Eccrine Angiomatous Hamartoma: A Clinicopathological Study of 26 Cases. Dermatology. 2015;231:63-9.
7. Patterson AT, Kumar MG, Bayliss SJ, Witman PM, Dehner LP, Gru AA. Eccrine Angiomatous Hamartoma: A Clinicopathologic Review of 18 Cases. Am J Dermatopathol. 2016 Jun;38:413-7.
8. García-Arpa M, Rodríguez-Vázquez M, Cortina-de la Calle P, Romero-Aguilera G, López-Pérez R. Multiple and familial eccrine angiomatous hamartoma. Acta Derm Venereol. 2005;85:355-7.
9. Castilla EA, Schwimer CJ, Bergfeld WF, Skacel M, Ormsby A. Eccrine angiomatous hamartoma in a neurofibromatosis type-1 patient. Pathology. 2002 Aug;34:378-80.
10. Shawa H, Hedayat A, Cotter D. Atypical presentation of pediatric antiphospholipid syndrome. JAAD Case Rep. 2021 Aug 18;16:50-2.
11. Lin YT, Chen CM, Yang CH, Chuang YH. Eccrine angiomatous hamartoma: a retrospective study of 15 cases. Chang Gung Med J. 2012 Mar-Apr;35:167-77.
12. Cebreiro C, Sánchez-Aguilar D, Gómez Centeno P, Fernández-Redondo V, Toribio J. Eccrine angiomatous hamartoma: report of seven cases. Clin Exp Dermatol. 1998 Nov;23:267-70.
13. Morrell DS, Ghali FE, Stahr BJ, McCauliffe DP. Eccrine angiomatous hamartoma: a report of symmetric and painful lesions of the wrists. Pediatr Dermatol. 2001 Mar-Apr;18:117-9.
14. Nunez MM, Selim MA, Flynn MS, Murray JC. Bilateral eccrine angiomatous hamartomas of the proximal interphalangeal joints. Dermatol Online J. 2023 Apr 15;29.
15. Tay YK, Sim CS. Eccrine angiomatous hamartoma associated with spontaneous regression. Pediatr Dermatol. 2006 Sep-Oct;23:516-7.
16. Kikusawa A, Oka M, Taguchi K, Shimizu H, Kunisada M, Nishigori C. Eccrine angiomatous hamartoma with sudden enlargement and pain in an adolescent girl after menarche. Dermatoendocrinol. 2011 Oct;3:266-8.