

Bilateral eccrine angiomatous hamartomas of the proximal interphalangeal joints

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Abstract

Eccrine angiomatous hamartoma is an uncommon, benign clinical entity constituting a nodular proliferation of eccrine glands and vascular structures localized to the dermis that typically present as unilateral, flesh-colored, erythematous, or violaceous papules on the extremities. These hamartomas may be associated with pain, hyperhidrosis, joint deformity, or functional impairment depending on the severity of the disease process. We present a case of bilaterally symmetric, asymptomatic eccrine angiomatous hamartomas involving all proximal interphalangeal joints of both hands. To date, there are only four prior cases of bilaterally symmetric eccrine angiomatous hamartomas reported in the literature, suggesting that the distribution experienced by our patient may represent a previously undescribed syndrome.

Keywords: angiomatous hamartoma, dermatopathology, eccrine glands, interphalangeal joint, proximal

Introduction

Eccrine angiomatous hamartoma (EAH) is a benign nodular proliferation of eccrine glands and vascular structures localized to the dermis. These lesions are often asymptomatic, though they are occasionally associated with a constellation of symptoms including hyperhidrosis and pain [1]. Eccrine angiomatous hamartoma classically presents in childhood on the extremities but can arise on other parts of the body including the face, neck, and trunk

with roughly similar prevalence in male and female patients [2-4]. Eccrine angiomatous hamartoma lesions are frequently unilateral; only four other published cases describing bilaterally-symmetric lesions exist to our knowledge [1,5,6]. These patients included a 13-year-old girl, 14-year-old boy, 14-year-old girl, and a 31-year-old man.

Case Synopsis

Herein, we describe a 63-year-old man with a dermatologic history notable for multiple atypical nevi who presented in 2010 with a 2-year history of skin-colored, non-tender papules on the dorsal proximal interphalangeal (PIP) joints bilaterally. The papules initially arose on the dorsum of his right second digit before gradually progressing to involve the right third and fourth digit as well as the left second, third, and fourth fingers. The papules were asymptomatic without joint swelling, hyperhidrosis, pain, or involvement of body regions other than the bilateral dorsal PIP joints.

On physical examination, the patient exhibited multiple skin-colored papules of up to 5mm in diameter. The papules became more prominent with flexion of the PIP joints and the skin moved freely over the lesions. Over the following years, the lesions gradually enlarged and eventually involved all five PIP joints bilaterally before stabilizing in size (**Figure 1**).

To rule out osteoarthropathies and other inflammatory processes, a generalized work up was



Figures 1. Eccrine angiomatous hamartomas (EAH) involving the proximal interphalangeal (PIP) joints. **A)** Bilaterally symmetric asymptomatic, skin-colored, papular lesions of all ten PIP joints, and **B)** a focused close-up of one lesion demonstrating no significant color changes of the overlying skin.

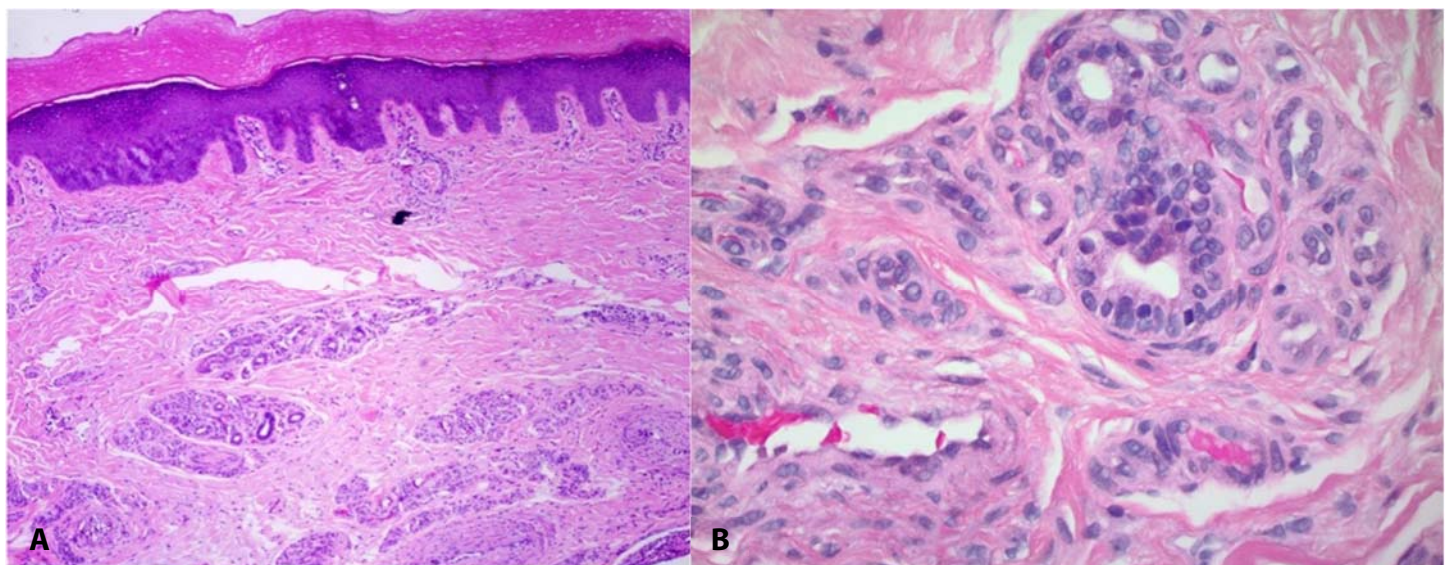
performed. An X-ray of the right hand demonstrated preservation of the joint spaces and was notable only for moderate first carpometacarpal joint osteoarthritis. Sedimentation rate, rheumatoid factor, and uric acid laboratory studies were within normal limits. Anti-nuclear antibody titer was 1:160. Histopathologic examination of a punch biopsy from the dorsal surface of the right fifth digit PIP joint revealed proliferative eccrine glands with blood vessels of various sizes in the middle and deep dermis with a normal epidermis (**Figure 2**). Taking the clinical presentation, histological findings, and laboratory data into account, the diagnosis of EAH was rendered. Since then, the patient has not reported development of any more papules

suspicious for EAH and his chronic EAHs remain asymptomatic.

Case Discussion

Eccrine angiomatous hamartomas typically present as a solitary nodule or plaque on the extremities with a broad range of coloration including erythematous, violaceous, or skin-colored but occasionally exhibit hues of blue, brown, or yellow. A few instances of multiple or bilaterally symmetric EAH, as seen in this patient, have been reported with an as yet unexplained preference for the extremities [1,2,5,6]. Skin biopsy with subsequent clinicopathologic correlation is diagnostic for EAH. Histologically, the proliferation of eccrine glands in the middle and lower dermis with associated ectatic or collapsed vessels with an overlying normal, acanthotic, or papillomatous epidermis are the hallmarks of EAH. The histopathology in this case was most consistent with this description.

Eccrine angiomatous hamartoma can be asymptomatic but are often associated with pain or hyperhidrosis secondary to eccrine glandular proliferation. Treatment can be offered for EAH if the lesions are associated with anatomic deformity of a joint, functional impairment, hyperhidrosis, or pain; sometimes treatment is desired for cosmetic reasons. Given this patient's asymptomatic course



Figures 2. H&E-stained section of punch biopsy of right fifth digit demonstrating a normal epidermis with abundant eccrine glands and associated vessels localized to the middle and deep dermis at **A)** low (10 \times), and **B)** high (40 \times) magnifications.

and lack of preference for cosmetic therapy, no treatment was warranted. However, analgesics for pain, botulinum toxin for hyperhidrosis, or surgical excision are conceivable options for symptomatic EAH [2,7].

Conclusion

We present an unusual distribution pattern of an already rare clinical entity. Only four cases of bilaterally symmetric EAH are reported in the literature to our knowledge. Only one of those patients was over the age of 18. Therefore, this case may represent a previously undescribed clinical syndrome, or perhaps a later-life manifestation of a syndrome of bilateral EAH with a clear predilection for the hands. Most recently, Clayton et al. described

a patient with bilaterally symmetric volar wrist nodules associated with hyperhidrosis identified as EAH following surgical excision [5]. The other three cited cases were localized to the dorsal rather than the volar surfaces of the wrists and fingers. Particularly of interest in our case is the exclusive involvement of the PIP joints. Our patient has not developed EAH on any other anatomic site since the diagnosis was established. We hope that our report may encourage others to provide the clinical and pathologic details associated with their own patients who have similar manifestations of EAH to further elucidate the clinical relevance.

Potential conflicts of interest

The authors declare no conflicts of interest.

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