CASE REPORT

Eccrine Angiomatous Hamartoma: A Report of 2 Cases

A. Batalla,a,* E. Rosón,a Á. Flórez,a A. Troncoso,b C. de la Torrea

aServicio de Dermatología, Complejo Hospitalario de Pontevedra, Pontevedra, Spain
bServicio de Anatomía Patológica, Complejo Hospitalario de Pontevedra, Spain

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Abstract Eccrine angiomatous hamartoma is a rare, benign tumor characterized by a proliferation of eccrine and vascular structures. We present 2 cases and review the characteristics of this disorder. The first patient was a 33-year-old woman who consulted for a brownish lesion on her back associated with local hyperhidrosis. The second patient was a 25-year-old man with an asymptomatic erythematous lesion on his left palm. In both patients a diagnosis of eccrine angiomatous hamartoma was made based on the histological findings.

Eccrine angiomatous hamartoma is usually present at birth or develops during childhood. It is typically a solitary lesion and signs and symptoms can vary; diagnosis is therefore based on histological study. The most common site is on the distal parts of limbs. The lesions tend to be asymptomatic, but there may be associated pain and hyperhidrosis. Treatment is not usually necessary except in cases with persistent symptoms, excessive sweating, or cosmetic concerns.

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KEYWORDS
Eccrine glands; Hamartoma; Hyperhidrosis

PALABRAS CLAVE
Glándulas ecrinas; Hamartoma; Hiperhidrosis

Hamartoma angiomatoso ecrino: presentación de dos casos

Resumen El hamartoma angiomatoso ecrino es un tumor benigno e infrecuente que se caracteriza por la proliferación de elementos ecrinos y vasculares. Presentamos dos casos, el de una mujer de 33 años que consultó por una lesión marronácea en la espalda, con hiperhidrosis asociada, y el de un varón de 25 años con una lesión eritematosa, asintomática, en la palma izquierda. Tras el estudio histológico ambos pacientes se diagnosticaron de hamartoma angiomatoso ecrino.

El hamartoma angiomatoso ecrino generalmente se presenta al nacimiento o en la infancia. Se manifiesta habitualmente como una lesión única con presentación clínica variable, por lo que su diagnóstico es fundamentalmente histológico. La localización más frecuente es la región distal de las extremidades. Suele ser asintomático, pero puede haber dolor e hiperhidrosis asociados. No requiere tratamiento salvo en aquellos casos asociados a sintomatología recalcitrante, sudor excesivo o por motivos estéticos.

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*Corresponding author.
E-mail address: anacebey@yahoo.es (A. Batalla).
Introduction

Eccrine angiomatous hamartoma (EAH) is a rare, benign tumor characterized by the proliferation of eccrine and vascular components.1,2 We present 2 cases of this hamartomatous tumor and review the characteristics of the disorder.

Case Descriptions

The first case of EAH was detected in a 33-year-old woman who consulted for a lesion on her back that had appeared 13 years earlier and was associated with local hyperhidrosis. There was no history of trauma or aggression in the area. Physical examination revealed a reddish-brown macule in the left dorsal region (Figure 1).

The second case occurred in a 25-year-old man who presented with an asymptomatic lesion on his left palm. It had appeared 17 years earlier and had grown progressively until the boy reached puberty, after which it stabilized. No association with possible triggers was identified. Physical examination revealed erythematous macules extending in a linear pattern from the tip of the middle finger to the center of the left palm (Figure 2).

Biopsy was performed in both patients. Histology showed hyperplasia of the eccrine glands closely associated with a proliferation of vascular channels and adjacent loose connective tissue (Figures 3 and 4). All of these findings were consistent with a diagnosis of EAH. The iodine-starch test showed hyperhidrosis in the area of both lesions (Figure 5). The first patient was treated with topical aluminium chloride, and the second patient received no treatment.

Discussion

The term eccrine angiomatous hamartoma was coined by Hyman et al in 1968, but the condition was probably first described by Lotbeck in 1959.1-5

EAH is caused by defective biochemical interactions between the differentiated epithelium and the underlying mesenchyma.1,3,5-8 It has also been described in association with radiation therapy, and a possible link between late-onset EAH and repeated trauma has been proposed.9,10

The true incidence of EAH is unknown and there is no difference between sexes.2,4-6,8,9,11,12 It appears before puberty in almost 70% of cases (49% are congenital), but it can also arise in adulthood.1,2,5-6,11 It typically manifests as a solitary lesion,1,4 ranging in size from 3 mm to 11 cm.1,2,8,9 It preferentially affects the limbs, and particularly the palmoplantar areas.13 The next most frequently affected sites are the trunk and the neck.1,5,6,8,11

It has heterogeneous clinical manifestations and can appear as plaques, nodules, or, albeit more rarely, papules or macules. The lesions can be red, reddish-blue, violet, yellow, brown, or pink.1,2,8,9 On occasions, there may be hypertrichosis.4

EAH tends to be asymptomatic, although it can present with pain or hyperhidrosis.1,2,5,13 Approximately 50% of patients experience pain, but this decreases in intensity
over the years. This pain is attributed to the presence of small nerves that penetrate the lesions. Several authors believe that pain and rapid lesion growth might be influenced by hormones as lesions can worsen during puberty and pregnancy. Hyperhidrosis, a manifestation of the eccrine component, occurs in a third of all patients with EAH and is seen in response to the typical triggers of eccrine secretion, such as heat or stress, although it can also occur spontaneously or after manipulation of the lesion. It is believed that the eccrine components are stimulated by the high temperature of the angiomatous component. There have been occasional reports of drug-induced or spontaneous resolution of hyperhidrosis.

Neither pain nor hyperhidrosis have been correlated with clinical presentation or lesion size.

Atypical clinical variants described in the literature include lesions that are similar to acanthosis nigricans and linear verrucous lesions and lesions that destroy the nail bed or cause osteolytic disorders. There have also been reports of concomitant knuckle pads, sebaceous nevus, and verrucous epidermal nevus. EAH is rarely associated with systemic disease, although there has been 1 report of EAH in association with neurofibromatosis type 1 and another in association with Cowden syndrome.

Typically, EAH is characterized by growth in parallel with the growth of the individual. Spontaneous regression is exceptional and there have been no reports of malignant transformation.

Histologic features include hyperplasia of normal or dilated eccrine sweat glands closely associated with angiomatous capillary channels and a variable presence of hair, lipomatous, mucinous, and lymphatic structures. The angiomatous component is characterized by thin-walled capillaries and dilated, thick-walled vessels within a dense fibrocollagenous stroma around the eccrine glands. Hamartomatous features of EAH include hair follicles, mucin, lymphatic structures, and even apocrine glands, adipose tissue, and dense connective tissue in the stroma. There are no mitotic or atypical cells. The lesions are located in the deep dermis and in subcutaneous cell tissue. The epidermis is usually normal, although there have been several reports of hyperkeratosis, papillomatosis, and acanthosis. Microscopically, the lesions are well circumscribed but they are not encapsulated. The immunohistochemical markers for EAH are similar to those used for normal eccrine glands.
The differential diagnosis for EAH is broad because of the wide spectrum of clinical variants of this disorder. This diagnosis should include vascular lesions, eccrine nevus, sudoriparous angioma, children's fibrous hamartoma, smooth muscle hamartoma, glomus tumor, Becker nevus, and angioleiomyoma. All of these conditions are histopathologically distinguishable.

Eccrine nevus and sudoriparous angioma are the 2 entities that are histologically most difficult to distinguish from EAH. Eccrine nevus, for example, is characterized by the proliferation of eccrine glands but there is no associated vascular component. One clinical finding that may help in the differential diagnosis is that hyperhidrosis occurs in the majority of cases of eccrine nevus but in just one third of those of EAH. The term sudoriparous angioma is surrounded by some controversy as it has been used synonymously with eccrine angiomatous hamartoma. It is also considered, however, that sudiparous angioma is a separate entity, distinguishable from EAH in that it has a more dominant angiomatous component and also contains dilated rather than hyperplastic eccrine elements.

Treatment is generally only necessary in cases of intense pain, excessive sweating, progressive growth, or cosmetic concerns. In such cases, surgery is the treatment of choice, but several authors recommend a watchful waiting approach if the surgery required is aggressive. An excisional biopsy may be curative for small tumors but more aggressive surgery may be needed for larger lesions, based on a preoperative assessment of tumor size, depth, and vascular supply. Ultrasound and nuclear magnetic resonance are valuable tools for this preoperative study. Other treatments include botulinum toxin, used to control excessive sweating, and polidocanol 1%, a sclerosing agent injected into the vascular component and that can lead to lesion regression.

In summary, EAH is a rare tumor that presents with a wide spectrum of nonspecific clinical features. The presence of pain or hyperhidrosis can help to raise suspicion of EAH, but histologic confirmation is required for a definitive diagnosis. Familiarity with the disorder and its good prognosis can help to reach a diagnosis and eliminate the need for unnecessary, aggressive surgery.

Conflict of Interest
The authors declare that they have no conflict of interest.

References