Multiple and Familial Eccrine Angiomatous Hamartoma

Mónica García-Arpa1, María Rodríguez-Vázquez2, Pilar Cortina-de la Calle1, Guillermo Romero-Aguilera1 and Rafael López-Pérez2

Departments of 1 Dermatology and 2 Pathology, Hospital del Carmen, Ronda del Carmen s/n, ES-13002, Ciudad Real, Spain. E-mail: monicagarpa73@mixmail.com
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Sir,

Eccrine angiomatous hamartoma (EAH) is a benign and uncommon combined vascular and eccrine malformation, often localized in the distal extremities. It may be congenital or appear later in childhood and rarely arises during adulthood. EAH may be solitary or more rarely multiple. Most of the time lesions are asymptomatic, although they may be painful and hyperhidrotic. We report three cases of multiple EAH seen at our department, two of them representing a mother and her son.

CASE REPORTS

Cases 1 and 2 (a boy and his mother): An 11-year-old boy had multiple 2–4 cm congenital and asymptomatic angioma-like plaques on the trunk and extremities which had been enlarging in size as he grew (Fig. 1). Moreover, he had more than six 1.5 cm cafe-au-lait macules on his shoulders and arms. He did not have a family history or other signs of neurofibromatosis (ophthalmological exploration did not reveal Lisch nodules and brain scan was normal). Histological examination of the angioma-like lesions revealed clusters of dilated vessels intimately associated with an increased number of mature eccrine glands, and with some adipose tissue lobules in dermis; which was consistent with EAH (Fig. 2).

After diagnosis of EAH in the boy, his mother, a 31-year-old woman, said that she had similar lesions since birth. She presented six asymptomatic and stable angioma-like plaques (1.5–3 cm in size) on her arms, hands and trunk. The biopsy of one angioma-like plaque showed proliferation of mature eccrine glands and capillary channels.

Case 3. One month later, a 2-year-old boy came to our department. He had 16 erythematous plaques measuring 0.5–2 cm on his right arm, right leg and trunk. These asymptomatic lesions had been present since birth and had been growing in size proportionally with growth of the patient. The biopsy specimen from one lesion showed a proliferation of ducts and lobules of eccrine sweat glands in the dermis consistent with EAH.

DISCUSSION

EAH was probably first described by Lotzbeck (1) in 1859, characterized histologically by tubular and glandular masses within a richly vascular stroma. In the 1960s, Vilanova et al. (2) described similar lesions as sweating angiomatous hamartoma and Domonkos & Suarez (3) described them as sudoriparous angioma. This was the same disorder that Hyman et al. (4) soon after described as EAH.

Fig. 1. Erythematous plaques on the trunk and left arm of case no. 1. These asymptomatic lesions had been present since birth.

Fig. 2. Histopathological findings of a biopsy from the elbow of case no. 1: increased numbers of well-differentiated eccrine sweat duct units and increased numbers of thin-walled vessels in mid and deep dermis (haematoxylin and eosin × 40).
To date, 47 cases of EAH have been reported in the literature, including our own three patients (4–16) (Table I). There is no sex preponderance. Nearly half the cases are congenital and 3/4 are present at puberty. They grow slowly and individual lesions range in size from 3 mm to 11 cm. Most cases (70%) are characterized by a single lesion. The clinical presentation of the lesions is very variable, ranging from a papule to a plaque even within a single patient. The colour may be red, yellow, brown or indistinguishable from normal skin. Most lesions appear on extremities, especially hands and feet (46% and 38%, respectively).

About one-third of all EAH (38%) lesions are asymptomatic. Pain and hyperhidrosis are reported together in 19% of cases. Pain alone is reported in a similar proportion and hyperhidrosis alone in 15% of patients. The tenderness of EAH has been attributed to small infiltrating nerves revealed by electron microscopy (5–7); however, some authors think that pain and rapid growth may be influenced by hormones because of onset or exacerbation during puberty and pregnancy. The lesional discomfort could be caused by fluid retention associated with pregnancy and menstruation (8). In general, this symptom disappears with excision of the lesion, but in some cases it may be necessary to amputate the toe or the finger to control pain (8). The focal hyperhidrosis may be an expression of its eccrine component (6, 9).

Histopathologically EAH shows a normal epidermis with occasional mild acanthosis. The dermis contains a proliferation of normal, sometimes enlarged, eccrine sweat glands in the mid and lower dermis intimately associated with thin-walled vessels, generally of capillary nature. Mitotic figures and cytological atypia have not been observed (10). Sometimes other elements have been found such as adipose tissue, pilar structures, lymphatics and mucin. The involvement of multiple structures indicates the hamartomatous nature of this skin lesion (10–12).

The immunohistochemical analysis of EAH has only been carried out in a few recent cases (11–14). As in normal eccrine glands, the secretory portion of the eccrine glands is positive for S100, carcinoembryogenic antigen (CEA), epithelial membrane antigen (EMA) and Cam5.2. The ductal components stained positively for CEA and cytokeratin 1 and weakly positive for EMA. The vascular elements labelled positively for anti-Ulex europaeus and anti-factor VIII antigens.

Clinically, EAH can mimic a capillary (‘strawberry’) angioma, vascular malformations (glomus tumour, blue rubber bleb naevus) and macular telangectatic mastocytosis. These lesions are differentiated by histopathological examination (10). Eccrine naevus and sudoriparous angioma are more difficult to differentiate. Most eccrine naevi show hyperhidrosis in contrast to approximately one-third of EAH. In sudoriparous angioma, the angiomatous component predominates and these vessels are predominantly large-calibre; eccrine glands are dilated but not hyperplastic (13).

The aetiology of this hamartoma is not clear. It has been suggested that it represents an abnormal induction of epithelial-adnexal and mesenchyme differentiation during early organogenesis (7). EAH lesions occur in acral skin where eccrine glands and vessels are generally numerous (11). However, other structures (adipose tissue, hair follicles, lymphatics) are also involved, suggesting an authentic hamartomatous nature (12).

EAH are benign lesions and no associated pathologies are usually reported; only one case of a 13-year-old boy with known familial neurofibromatosis type-1 and one congenital EAH is reported (14).

This entity usually does not require treatment except for aesthetic reasons. Lee et al. (15) reported two patients with symptomatic EAH (pain and hyperhidrosis) who were treated with a flash-pumped-pulsed dye-laser with variable results. Botulinum toxin might be considered in hyperhidrotic cases.

**REFERENCES**


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**Table I. Clinical features of 47 cases of eccrine angiomatous hamartoma reported in the literature.**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>n (%)</th>
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<tbody>
<tr>
<td>Age at onset</td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>23 (48.9)</td>
</tr>
<tr>
<td>Prepubertal</td>
<td>11 (23.4)</td>
</tr>
<tr>
<td>Adult</td>
<td>11 (23.4)</td>
</tr>
<tr>
<td>Number of lesions</td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>33 (70.2)</td>
</tr>
<tr>
<td>Multiple</td>
<td>14 (29.8)</td>
</tr>
<tr>
<td>Location</td>
<td></td>
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<tr>
<td>Extremities</td>
<td>39 (83)</td>
</tr>
<tr>
<td>Trunk</td>
<td>12 (25.5)</td>
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<tr>
<td>Neck</td>
<td>1 (2.1)</td>
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<tr>
<td>Clinical structure</td>
<td></td>
</tr>
<tr>
<td>Plaque/macule</td>
<td>25 (53.2)</td>
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<tr>
<td>Nodule</td>
<td>17 (36.2)</td>
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<tr>
<td>Papule</td>
<td>5 (10.6)</td>
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