Eccrine angiomatous hamartoma: case report and clinical, pathologic and ultrasonographic studies

Hamartoma angiomatoso écrino: relato de caso e estudo clinicopatológico e ultra-sonográfico

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Abstract: Eccrine angiomatous hamartoma is an unusual neoplasia characterized by hamartomatous proliferation of the portion of the secretory and ductal sudoriparous eccrine gland associated with capillary and other elements. We characterized the clinicopathologic and ultrasonographic study of a bluish nodule in the digital pulp of the right hand of a nineteen-year-old girl. The nodule was painful and associated with sweating.

Keywords: Hamartoma; Hemangioma; Ultrasonography

INTRODUCTION

Eccrine angiomatous hamartoma (EAH) is a rare benign neoplasia characterized by proliferation of the secretory portions of the eccrine sudoriparous glands associated with capillary angiomatosis and proliferation in other elements, such as in the adipose tissue, hair and epidermis.

Most cases occur at birth or in early childhood, with various forms of clinical presentation, ranging from simple angiomatous nodules to erythematous-purpuric plaques. Multiple lesions may occur, however the predominant picture is of a solitary lesion.

AEH is also known as sudoriparous angioma when there is a prevalence of angiomatous elements and when the eccrine elements are dilated, but not hyperplastic. However some authors consider these to be separate entities.

This work describes the clinicopathologic and ultrasound characteristics of a patient with a typical lesion of this disease.
CASE REPORT

The patient was a 19-year-old woman, who had a bluish nodule in the digital pulp of the right hand, with onset two years before. The lesion was pruriginous, slightly painful upon compression and associated with sudoresis. The patient denied having any comorbidities or allergies and also denied the use of medications and similar cases in the family.

At physical examination, she was in good general condition, good color, eutrophic and without alterations, except for the presence of the violaceous nodule in the digital pulp of the right hand. The lesion was well circumscribed, with a diameter of 2 cm (Figure 1), slightly painful upon compression and which presented hyperhidrosis that was confirmed by a positive reaction in the iodine and starch test (Figure 2).

Ultrasound exam was done using a 7.5 MHz transducer, revealing a homogeneous, well-circumscribed tumor, located in the fatty tissue and with vascularization of moderate intensity detected by Doppler ultrasonography (Figure 3).

Surgical excision was performed revealing a tissue of fibroelastic consistence and a violaceous coloration.

The histopathologic analysis of the sample stained by hematoxylin-eosin showed dilated blood vessels and a significant number of eccrine sudoriparous glands. (Figure 4).

DISCUSSION

Lotzbeck, in 1859, described an angiomatous lesion in a child that was located above the malar region, and was characterized histologically by glandular structures amidst the vascular stroma. In 1895, Bier described a case of pigmented angioma associated with spontaneous pain and hyperhidrosis in the lesion. Fine (1961) described the same type of lesion in an adolescent, as "blue rubber bleb nevus", with multiple bluish nodules, pain and local transpiration. The patient did not exhibit gastrointestinal lesions or bleeding. Finally, Hyman et al. (1968) used the expression eccrine angiomatous hamartoma (EAH) to encompass all the names found in the literature referring to this lesion, such as, for instance, angiomatous hamartoma with functional transpiration, hangiomatous hamartoma with secretion of perspiration and angioma with nevus of a sudoriparous gland.

EAH presents clinical characteristics that are identified by vascular proliferation much more than by the increase in the number of sudoriparous glands. However, there are reports about the existence of other components, especially pilose follicles.

EAH occurs with an equal incidence between both sexes, with approximately a third of the cases manifesting at birth or in early childhood. Few references exist about lesions that have appeared in adult life or in puberty, such as is described in this case report.

Clinical manifestations may vary from nodules to plaques of erythematous-bluish or brownish color. They may be located in any part of the body, although they are more frequently found in the palmar regions, such as in the patient herein described.
It is commonly associated with hyperhidrosis and pain that is spontaneous or follows local pressure. Probably the pain occurs due to involvement of nerve fibers, and hyperhidrosis because of the stimulation of the eccrine components, caused by the elevated local temperature within the angioma. The differential diagnosis includes: vascular malformations, hamartoma of the smooth muscles, glomus tumor, mastocytosis, telangiectasia macularis and tufted angioma. All of which are differentiated by their histopathologic and immunohistochemical characteristics.

Immunohistochemical study (not performed in this case) can demonstrate that antigens frequently found in eccrine glands, such as the carcinoembryonic antigen and protein S-100, are reduced qualitatively in the eccrine glands of EAH, while *Ulex europaeus*, CD34, CD44 and the antigen related to factor VIII are expressed by the endothelial cells of the vascular components.

The treatment for small lesions is surgical. Extensive or asymptomatic lesions can receive only corrective treatment. The present study describes the typical clinical, histopathological and ultrasonographic, characteristics compatible with the described case.
REFERENCES


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