Ultrasound Study of Eccrine Angiomatous Hamartoma: Two Cases Report

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Abstract

The Angiomatosis Ecrine Hamartoma (EAH) is a benign and rare tumor, located in areas with abundant eccrine glands (extremities-acral regions). Its clinical presentation can be diverse: papule, plaque, nodule or tumor. The Pathological Anatomy is characterized by the proliferation of mature eccrine glands and dialatal vascular channels in the dermis. At present there are no sonographic descriptions of EAH.

Keywords: Dermatological ultrasound; Eccrine glands; Benign tumors of the eccrine glands; Eccrine angiomatous hamartoma

Introduction

Hamartomas can be grouped in pure (pure eccrine nevus and eccrine hamartoma) and mixed, the varieties include angiomatous component, mucinous and lipomatous [1].

Eccrine angiomatous hamartoma is an exceedingly rare, benign tumor that usually exhibits from birth, congenitally or is acquired during childhood [2-4]. Its most frequent location is on extremities [3-6], although there are published cases of EAH on the trunk [7]. The lesions are clinically variable: nodules, plaques, papules or tumors that vary in color between red, yellow, bluish, brown or combination of both. They usually appear as single lesions [3]. The EAH can either be asymptomatic or be linked to pain or hyperhidrosis. It is thought that the pain on the lesions is secondary to the infiltration of small nerves from the epidermis [5].

The histological examination of EAH is characteristic, as it presents intradermal proliferation of mature eccrine glands and dilated vascular channels [8].

The clinical differential diagnosis includes hemangioma of infancy, eccrine nevus and tufted angiomia amongst others.

Series of Cases

Clinical case 1

An 8-year-old boy, who presented with a semi-soft, non-adherent and painless on palpation nodule on the back of the left foot, close to the external malleolus. It was well-defined, skin-colored with some brownish areas, and it had over a year of evolution. On the right foot, a bilateral, symmetrical, incipient lesion with similar characteristics was observed. The child presented with valgus attitude of the feet (Figure 1).

Figure 1 Valgus attitude of the feet.

The ultrasound study in mode B, with linear probe of 18 MHZ, showed an epidermis with increased thickness and echogenicity. In the deep dermis, there is a well-defined area, 16 x 4 mm in diameter, homogeneous, hypoeochic, with small anechoic areas in its interior, which are evident in salmon color (Figure 2), the doppler came out negative. The anechoic zones inside the lesion can be observed with a higher magnification.
The cutaneous biopsy showed a hyperkeratotic epidermis. In the deep dermis, some glomerular structures were identified. They were composed of a double component: arteriovenous vessels and another ductular component of fully mature eccrine sweat glands (Figure 3). The diagnosis was eccrine angiomatous hamartoma. The lesion was partially excised. The remaining hamartoma continued to cause a thickened appearance on both of the affected fingers.

Sonographic evaluation was performed to follow up with a linear probe with variable frequency up to 18 MHz. Gray scale imaging showed a dermal and hypodermal heterogeneous area, mostly hypoechoic, with small anechoic lacunar areas contained therein. Doppler color mode did not reveal any increased vascularity (Figure 4).

**Figure 2 Ultrasound study in mode B.**

**Figure 3 Biopsy showing increased number of blood vessels and eccrine glands.**

**Figure 4 Sonographic evaluation.**

**Discussion**

Lotzbeck first described EAH in 1859. It was in 1968 when Hyman et al. gave it its current name.

It is a benign and rare tumor, with no predilection for sex, nor a characteristic inheritance pattern. Although there have been reports of causes related to Neurofibromatosis I and another one to Cowden Syndrome, it is not associated with systemic alterations [9]. The growth of EAH may be due to hormonal factors - such as puberty, repeated trauma [10] - which could explain the appearance and growth in our first case, where the patient had a valgus attitude more marked on the left foot which repeatedly produced traumatisms because of friction with the boots.

EAH has several clinical presentations, such as a macula, plaque, yellow-brown nodule - as shown in our first case, or skin-colored - as in our second case.

The two cases presented coincide with the most frequent locations of EAH presentation, which are the acral regions (extremities) [11].

Approximately, 40% of EAH cases can be linked to hyperhidrosis, which gives the appearance of an "angioma that cries" [12].

The treatment of EAH will be done by taking into account: the age of the patient, the size and especially the symptomatology that is present [13]. In small or asymptomatic lesions, the behavior can be expectant. If the injury hurts, the treatment of choice is surgery. In lesions with significant

**Clinical case 2**

A 14-year-old boy who displayed dermal thickening on the second and third finger of the left hand since birth (Figure 1).

A biopsy performed at the age of 6 revealed a dermis and a hypodermis with an increased number of blood vessels and eccrine glands (Figure 3). The diagnosis was eccrine angiomatous hamartoma. The lesion was partially excised. The remaining hamartoma continued to cause a thickened appearance on both of the affected fingers.
hyperhidrosis, there is record of cases treated with good results using botulinum toxin [14]. Spontaneous regression is infrequent, but it has been reported [15].

In clinical case number 1, the patient was referred to the Rehabilitation service to correct the valgus posture of his feet. The smaller lesion of the right foot completely disappeared and the one on the left foot decreased considerably in size. In clinical case 2, the lesion was completely intervened, for which the use of dermatological ultrasound was very useful, since it helped calculate the size of the persistent lesion.

The ultrasonographic characteristics collected in our two clinical cases are:

- Hypoechoic areas, regular borders, oval morphology, well-defined, located in the deep dermis.
- Anechoic areas within the lesion, which could correspond to the ductal component of mature eccrine sweat glands (Figure 5).

Figure 5 Ductal component of mature eccrine sweat glands.

The color Doppler with linear probe of 18 MHz was negative in both cases, perhaps because of the frequency of the probe or the vascular component with microscopic characteristics, which allowed us to perform a differential diagnosis with lesions of vascular origin such as infantile hemangioma.

Parra et al. in a review of the literature and presentation of nine cases of EAH, coincides as our patients in that the most frequent location is the extremities and that some cases are caused by repeated trauma, as occurred in our first patient.

In 2005, Costa et al. described the ultrasonographic study about an Eccrine angiomatous hamartoma’s case. In her paper She described: “The Ultrasonography is safe, non-invasive, used in the diagnosis of tumors of tissues moles. The examination does not allow histological diagnosis to differentiate between benign and malignant tumors. The Ultrasound characteristic combined with experience of the examiner and the clinical aspects allows a more specific diagnosis. The Doppler flowmeter in this case demonstrated blood vessels of uniform shape”. A di difference with our cases with negative Doppler.

Currently, there are much more precise, precise and less restrictive imaging techniques than ultrasound for the diagnosis of vascular malformations such as:

- MRI: represents the best diagnostic method in assessment of vascular anomalies as regard to their soft-tissue components and relationships with nearby structures.
- Conventional MRA techniques, such as time-of-flight, phase-contrast MRA and contrast enhanced MRA, may allow useful information about feeding vessels of lesions, but they produce static images with prolonged acquisition time and cannot detect small vessels.

Time-resolved imaging of contrast kinetics (TRICKS) is a recently introduced technique of contrast enhanced MRA that acquires multiple 3D volumes during the passage of contrast agent bolus, so one can obtain dynamic filling of the arteries and veins similar to digital subtraction angiography (DSA) [16-19]. Many studies have demonstrated the advantage of this technique in assessment of the peripheral, intra- and extracranial as well as spinal vascular lesions [20].

Conclusion

We believe that dermatological high frequency ultrasound with color Doppler is a tool that can be used for the diagnostic orientation of skin lesions that appear during childhood.

The importance of these cases lies in the fact that they are the first ones in the reviewed medical bibliography literature to be described ecographically with 18 MHz.

References


