CONGENITAL SMOOTH MUSCLE HAMARTOMA. COMMENTS ON A CASE

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Summary

Congenital smooth muscle hamartoma is an uncommon lesion of a dysembryoplastic nature. Most often, it is present from birth, as a single plaque. The rubbing of the lesion can lead to the appearance of the Darier's pseudo-sign. The form of congenital smooth muscle hamartoma manifested as a single plaque is benign and it is not associated with other pathologies. We present the case of a 6-month-old child, consulted for a brown-pink plaque, located on the left thigh.

Keywords: *congenital smooth, clinical, histopathology.*

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Introduction

Congenital smooth muscle hamartoma (CSMH) or leiomiomatous hamartoma is an uncommon lesion of a dysembryoplastic nature. The name origin is Greek language: hamartia = defect, mistake, error; and *-oma* = tumor/ neoplasm. The first case of CSMH was described by Sourreil et al. in 1969 [1]. The most common CSMH shows itself as a single plaque, being present from birth or manifesting during the early infancy period. Rubbing the lesion can lead to the appearance of the Darier's pseudo-sign as a transient induration and piloerection.

Clinical case

The 6-month-old child, born at term, was consulted for a slightly hardened plaque, measuring 6 by 4 cm, with a brown-pinkish color, covered with black hairs and located on the left thigh (Fig. 1). The plaque appeared from birth. The pseudonym Darier was not present. The

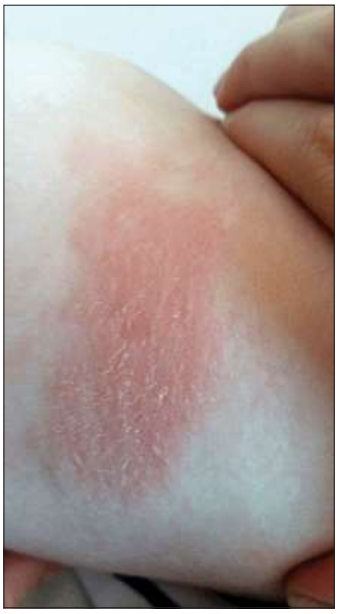

Figure 1. Clinical appearance in 6-month-old children: plaque with hypertrichosis on left thigh

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pregnancy went normally, and the birth was without complications.

Histopathological examination showed the presence in the dermis of numerous bundles of hyperplastic smooth muscle fibers of normal morphology, anarchically distributed. The epidermis did not show significant histological changes (Fig. 2). The diagnosis of congenital smooth muscle hamartoma was established.

Figure 2. Microscopic appearance of the hamartoma section: hyperplastic smooth muscle fibers, with various sizes and anarchic orientation, distributed in the dermis (H&E, x12)

Discussions

CSMH is a rare skin hamartoma (1/2700 newborns), developed from the erector muscles of the hair and it is usually present from birth. Clinically it presents it self as a fixed plaque which can be slightly infiltrated or not, with dimensions of a few centimeters, with or without follicular papules on its surface, asymptomatic or discreetly itchy. Hyperpigmentation is observed in more than half of the cases, but the most common and at the same time embarrassing sign is hyperpilosity. The hairs are long, black and more visible than those of the adjacent healthy skin. In half of the cases, the rubbing of the plaque leads to the appearance of Darier's pseudo-sign, which is manifested as erythema and transient piloerection [2]. The favorite locations are the lumbar region, the arms, the thighs, the buttocks and exceptionally the flanks, the face, the hairy scalp and the mammary region.

Histopathological examination reveals the presence in the dermis of bundles of smooth muscle fibers of normal morphology but in large numbers, arranged disorderly, at a variable distance from hair follicles, separated by collagen fibers, under a discrete hyperplastic epidermis [3,4].

The most common CSMH is presented as a single plaque, but there are rare cases of CSMH with plaques involving multiple sites. In some cases, the form of CSMH in multiple plaques can give the appearance of a Michelin baby, an aspect that can also be given by the lipomatous hamartoma. The form of CSMH in a single plaque is benign and can sometimes have an involutive evolution, never being associated with other pathologies [5]. The intermediate multiple plaque clinical form has a good prognosis as opposed to the Michelin baby clinical type which is most often associated with neurological or osteo-articular pathology.

The differential diagnosis of CSMH includes conditions with hyperpilosity at birth. In the localized form, differential diagnosis is made with congenital nevocellular nevus, nevoidic hypertrichosis, and with spinal dysraphism in certain localizations [6]. In the clinical form of CSMH with multiple plaques, should be eliminated fetal alcohol syndrome, fetal hydantoin syndrome and complex malformative syndromes. The acquired forms of hypertrichosis, which appeared on a pigmented plaque, bring into discussion the muscular hamartomas with late onset and the Becker's hamartoma [7,8]. Becker's hamartoma differs from CSMH by age of onset, morphological aspects with more obvious epidermal changes (hypertrichosis and hyperpigmentation) and histopathological changes as lower development of smooth muscle fibers.

Conclusions

CSMH is a rare, disembryoplastic condition that usually occurs from birth as a single plaque. The most common differential diagnosis is Becker's nevus, with peripubertal-onset.

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Conflict of interest
NONE DECLARED

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