CASE REPORT

Congenital Smooth Muscle Hamartoma. Report of a case and brief review of the literature.

Verónica Uraga,* Cristina Solorzano,* Gabriela Crespo,* Juan Carlos Garcés, Enrique Uraga*****

* Dermatologists from the Dermatological Center "Dr. Uraga"
** Dermatopathologist
*** Director of the Dermatological Center "Dr. Uraga"

Correspondence:
veronica_uraga@hotmail.com

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ABSTRACT

Congenital hamartoma of smooth muscle is a benign proliferation, generally solitary of smooth muscle, quite rare and predominantly in males. It is usually located in the trunk or in the root of the limbs with variable degree of pigmentation and hypertrichosis, sometimes presenting positivity of the so called pseudo Darier's sign. We present the case of a 2 year old girl with a lesion on her left knee present since birth and we make a small review of the literature on the subject.

INTRODUCTION

Congenital smooth muscle hamartoma (SCML), also known by the names arrector pili hamartoma and pilar smooth muscle hamartoma, is a benign proliferation described by Stoke et al. at the beginning of the 20th century, of relatively rare observation, with an incidence of 1 in 2600 births[1] and with a slight predominance in males. It presents at birth in the form of a skin-colored or slightly hyperpigmented patch or plaque, pigmentation that increases in adults. Prominent hair can also be seen. When there is an increase in hair, the number of hairs is not increased, but there is an increase in their length and diameter.[2] Other authors speculate that there is a relationship between hypertrichosis, including hair density, and the amount of smooth muscle bundles.[3] It is generally a single lesion and its preferential, but not exclusive, locations are the trunk and the roots of the extremities.[4] Rubbing of the lesion frequently leads to contraction of the smooth muscle due to stimulation of the arrector pili muscles, all of which determine elevation and transient induration and piloerection giving rise to the pseudo Darier sign, reminiscent of the Darier sign that is It is produced by the release of histamine from mast cells.[5] This sign has been reported atypically, presenting the induration in the form of cobblestones.[6] Rubbing of the lesion frequently leads to contraction of the smooth muscle due to stimulation of the arrector pili muscles, all of which determine elevation and transient induration and piloerection giving rise to the pseudo Darier sign, reminiscent of the Darier sign that is It is produced by the release of histamine from mast cells.[5] This sign has been reported atypically, presenting the induration in the form of cobblestones.[6] Rubbing of the lesion frequently leads to contraction of the smooth muscle due to stimulation of the arrector pili muscles, all of which determine elevation and transient induration and piloerection giving rise to the pseudo Darier sign, reminiscent of the Darier sign that is It is produced by the release of histamine from mast cells.[5] This sign has been reported atypically, presenting the induration in the form of cobblestones.[6]

The histopathological study reveals a random proliferation of smooth muscle bundles in the reticular dermis without atypia or inflammatory activity. If necessary, immunohistochemical tests can be performed that reveal smooth muscle positivity for actin.[7]

CASE REPORT

This is a two-year-old female patient who consulted due to presenting an injury to her left knee from birth. The mother reports that she started as a small papule of 1.5 cm x 1.5 cm that later grew slowly. At the time of the examination, a plaque with a size of 5 cm x 6 cm was observed, somewhat infiltrated to the touch, normal skin color, with a discreet presence of hair. (Photo 1)

With the presumptive diagnosis of HCML we rubbed the lesion and observed thickening and infiltration (Photo 2) as well as piloerection (Photo 3) which we classify as Darier's pseudo sign.

With this diagnosis, we performed a punch biopsy and it was sent for histopathological study, which reported:

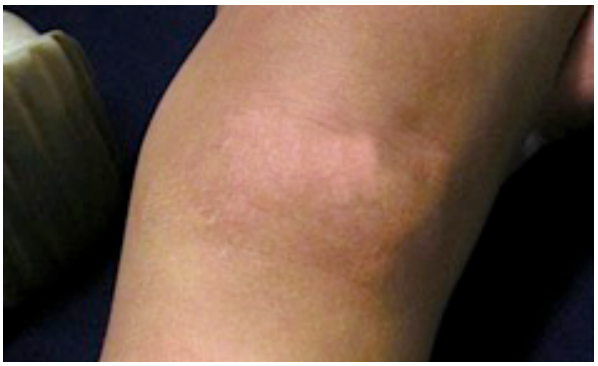

Foto 1. Lesión en rodilla izquierda, algo elevada levemente rosada.

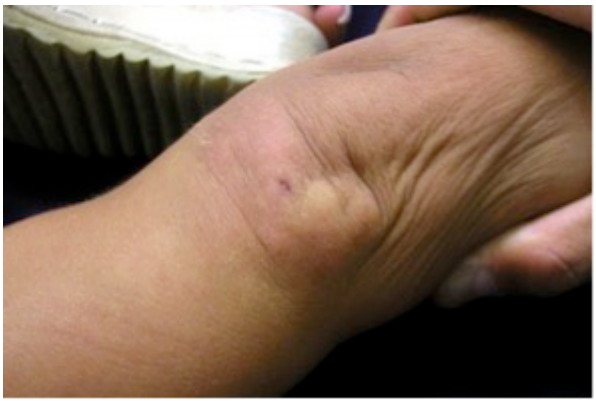

Foto 2. Pseudo signo de Darier pos frotamiento de la lesión.

Histological sections show skin with mild hyperkeratosis and granulosa enhancement. The dermis shows proliferation of collagen fibers. A hair follicle and arrector pili muscles are seen in an irregular arrangement. The proliferation of collagenous fibers deepens into the hypodermis being in contact with the edge of the section. (Photos 4-5)

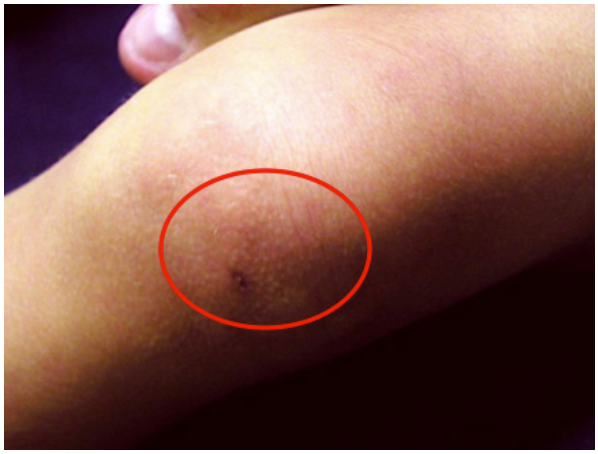

Foto 3. Pseudo signo de Darier. Se observa piloerección (círculo pos frotamiento).

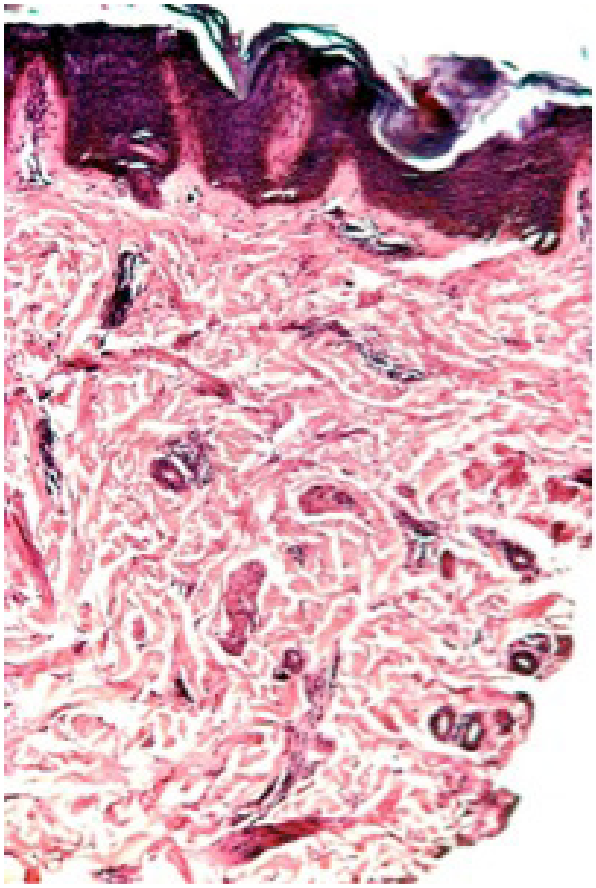

Foto 4. Histopatología que permite observar las alteraciones reportadas anteriormente.

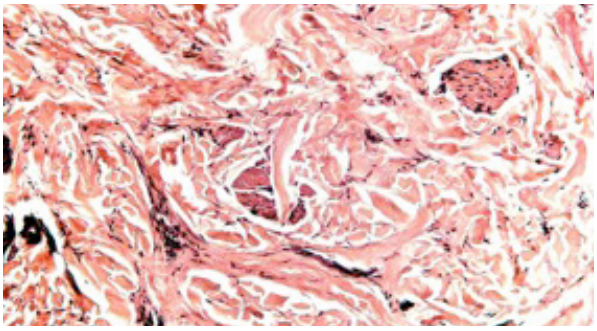

Foto 5. Histopatología anterior con mayor aumento.

DIAGNOSIS

Smooth Muscle Hamartoma

CONCLUSION

HCML is a congenital or acquired condition, whose diagnosis is essentially clinical and histopathological, although dermoscopy currently appears as support, especially in visualizing hairs and prominent follicular openings and to check the same aspects after rubbing to observe the pseudo sign. Darier's.[8] We have previously indicated that the preferential location sites were the trunk and the root of the limbs, however, other areas of appearance are reported, such as the tongue,[9] face,[10] ankle,[11] scalp.[12]

The clinical forms of HCML include the classic one, which we have already mentioned, the papulofollicular form that presents with multiple skin-colored papules that can unite to form irregularly shaped plaques, and finally, a mixed form combining the two previous ones.[11] It is generally a single lesion but multiple locations are reported.[13] Usually their size ranges between 2 and 5 cm, however sometimes they can reach diameters that reach 15-20 cm[14] or even gigantic.[15]

We report this case because it seems to us that it is a pathology of not so frequent observation and whose proper knowledge allows a clinical diagnosis and its final histopathological verification.

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