CASE REPORT

Hairy nevus lipomatosus cutaneous superficialis: A rare presentation

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INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare idiopathic hamartomatous condition that is histopathologically defined by the presence of ectopic mature adipose tissue in the dermis. Hoffmann and Zurhelle were the first to describe it in 1921. Since then, 2 main clinical variants have been described. The classic type is considered the most common form and clinically presents as multiple, grouped, pedunculated, cerebriform nodules that are yellowish or skin colored. They can coalesce to form a large plaque and are distributed in a zosteriform pattern. The second type is the solitary form, which presents as a solitary dome-shaped sessile papule or nodule with a nonspecific distribution.

However, multiple reports have also described different clinical presentations, such as giant NLCS, NLCS with multiple open comedones, and NLCS with ulcerations and necrosis. In 1972, Finley and Musso described increased hair growth over the nevus. However, to our knowledge no other reports have described this clinical variant. Therefore, we report a case of a child with NLCS, which is notable because of its rarity and unusual presentation with hypertrichosis overlying the NLCS.

CASE REPORT

An 8-year-old boy with no relevant medical history presented with a slowly growing lesion over his right ankle, which he had had since birth (Fig 1). In a few months, the lesion started to elevate and grew dark coarse hair. Later, similar lesions developed around the original one. The lesions were asymptomatic, with no history of ulceration or discharge. They did not affect his mobility. He had normal development and achieved all milestones at an appropriate age. His family history was unremarkable, with no history of similar lesions.

Histopathologic examination revealed mature adipose tissue in the dermis (Fig 2). The clinical findings, along with the histologic features, indicated a diagnosis of NLCS.

DISCUSSION

Nevus lipomatosus is a rare idiopathic hamartomatous anomaly that is characterized by 2 clinical entities. The classic form usually presents during the first 3 decades of life and is characterized by multiple soft yellow to skin-colored, multilobulated, plaques with overlying terminal hypertrichosis. The plaques had an arcuate distribution around the ankle and measured 2.5 × 1 cm, 2.5 × 3 cm, and 2 × 1 cm. There were no associated café-au-lait macules or hypertrichosis in other areas. Palpation of the lesion did not provoke any symptoms, and the rest of the physical examination result was unremarkable. Histopathologic examination revealed mature adipose tissue in the dermis (Fig 2). The clinical findings, along with the histologic features, indicated a diagnosis of NLCS.

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type usually appears during the third to sixth decades of life as a single, soft, yellowish to skin-colored nodule, which can occur on any location on the skin, including the scalp, eyelids, and clitoris. In our patient, the onset of the lesion, the site, and the clinical appearance were consistent with the classic form. Lesions are asymptomatic in both forms, as observed in our patient. Various overlying skin changes have been reported, including ulceration, particularly after trauma or ischemia; café-au-lait macules; leukodermic spots; and comedolike alterations. However, the presence of hypertrichosis, as in our patient, is a unique feature. To the best of our knowledge, this feature has been reported only once, in 1972 by Finley and Musso.

The histopathologic features in our patient revealed basket-weave hyperkeratosis of the epidermis with underlying infiltration of the dermis by mature and unremarkable fat cells, which compromised approximately 70% of the dermis (Fig 2). Adipocytes were aggregated around blood vessels and eccrine glands. These findings are similar to previous observations. Two unremarkable hair follicles were also observed (Fig 3).

Elastic Van Gieson stain showed normal dermal elastic fibers (Fig 4).

An important differential diagnosis that is clinically similar to NLCS is smooth muscle hamartoma, which shares the feature of plaque with hypertrichosis, similar to the presentation of our patient. However, histologic examination was sufficient to differentiate between these 2 conditions and confirm the diagnosis of NLCS.

We have reported a case of hairy NLCS to highlight its rarity and its clinical similarity to smooth muscle hamartoma, as well as to emphasize the need for histologic examination for accurate diagnosis.

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