



NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS- A RARE PRESENTATION

Dermatology

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ABSTRACT

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous skin tumor characterized by dermal deposition of mature adipose tissue. It was first described by Hoffmann and Zurhelle (1) in 1921. Clinically it is classified into two clinical subtypes: a multiple form and a solitary form. The lesions are usually found in the flank, buttocks, and upper part of the posterior aspect of the thigh, but they can occur on the abdomen, chest, and face.(2) We report a case of NLCS present in the left axillary region because of its rarity and its unusual site.

KEYWORDS

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign cutaneous hamartoma. It was first described in 1921 by Hoffman and Zurhelle.⁽¹⁾ It is defined by the presence of mature adipocytes among the collagen bundles of the dermis.^(2,3) This condition is usually present at birth or in childhood with no familial tendency nor sex predilection.^(4,5) Clinically, it is classified into two clinical variants, the classical and the solitary form. The classical type is usually composed of multiple and grouped skin-colored, pedunculated nodules. A second type is characterized by a solitary dome-shaped sessile papule or nodule.⁽⁶⁾

Although fat deposition in the dermis has been considered to be a consequence of degenerative changes in connective tissues, the pathogenesis of NLCS remains unknown.⁽⁷⁾ Several theories have been proposed. One of them suggests that multiple type NLCS arises from adipose metaplasia in the course of degenerative changes in the dermal connective tissue. Another theory is that the adipocytes would represent a true nevus from the developmental displacement of adipose tissue.⁽⁸⁾ A third proposal is that the mature adipocytes grow from mononuclear cells differentiating into lipoblasts in a perivascular area.⁽⁹⁾

The histopathology of NLCS shows clusters of ectopic mature adipose tissue among collagen fibers in the dermis with no connection to the subcutaneous fat tissue.⁽²⁾ These ectopic adipocytes contain large intracytoplasmic lipid vacuoles and are often associated with vascular structures.⁽⁹⁾

We described an unusual case of an NLCS present since birth in a 25-year-old male patient.

Case Report

A 25-year-old unmarried male came to us with complaint of multiple asymptomatic lesions over his left underarm since birth which had gradually increased in size over a period of 25 years. There were no systemic complaints nor any familial history of similar lesions present. On clinical examination, multiple, well-defined, discrete and confluent, skin colored to yellowish, soft, sessile nodules, and plaques were seen.(fig 1) Few lesions showed cerebriform surface and few lesions were hyperpigmented due to friction. Multiple skin tags were also present. There was no ulceration, discharge, excessive hair growth, comedo like plugs or cafe-au-lait spots. An incisional biopsy was performed from the papule which revealed islands of mature adipose tissue in the dermis. Based on the clinical evaluation and histopathological findings, the diagnosis of nevus lipomatosus cutaneous superficialis was confirmed. Later the patient was referred to surgery department for further management.



DISCUSSION

NLCS is a rare idiopathic benign hamartomatous anomaly that is characterized by ectopically situated mature adipocytes in the dermis.^(4,7) It may be present at birth or early childhood. It is classified into two clinical forms. The classical or the most common form is characterized by clusters of soft, skin to yellow colored papules or nodules, with smooth and wrinkled appearance, located on the lower back, pelvic girdle, buttocks and upper thighs in a zosteriform pattern.⁽⁷⁾ It appears during the first three decades of life. The solitary or the rare type can occur anywhere on the skin and usually appears as a single nodule, with the same dermatologic appearance.⁽⁴⁾ This clinical form has been noted in rare sites such as the face, scalp, eyelids, and clitoris.^(2,7) It appears usually during the third to the sixth decade of life. In our case, the patient had multiple, cluster of soft, skin colored papules and nodules over his left underarm since birth and the dermatological characteristics were compatible with the classical form.

In both types, the lesions are asymptomatic and invites concern when there is aesthetic issue, as seen in our patient. Ulceration occurs rarely, mostly due to external traumas or ischemia.^(2,7) Café-au-lait macules, leukodermic spots, overlying hypertrichosis and comedo-like alterations can coexist.^(3,7)

The histopathology of NLCS usually reveals proliferation of ectopic mature adipocytes in the reticular dermis that varies from 10-50% of the lesion.^(5,10) The adipocytes most commonly form small aggregates around blood vessels or eccrine glands, but may also be present as solitary adipocytes between collagen bundles.⁽¹²⁾ In some cases, there is an increased density of collagen fibers, fibroblasts, and a perivascular infiltration of mononuclear and spindle-shaped cells.^(7,10) The epidermis exhibits acanthosis, basket weave hyperkeratosis, increased basal pigmentation, and obliteration with focal elongation of rete ridges. Adnexal structures may be unaffected or reduced in some cases, and may show perifollicular fibrosis.⁽¹⁰⁾ In our patient, the histological findings were consistent with NLCS.

Clinically, NLCS should be differentiated from nevus sebaceous, connective tissue nevus, neurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (Goltz syndrome).^(2,7) Histopathological studies usually help in differentiation and a biopsy should be performed to confirm diagnosis. Similar dermal collections of adipocytes are also present in some melanocytic nevi, pedunculated lipo-fibromas, and in Goltz syndrome. In the case of Goltz syndrome, besides the clusters of adipocytes in the dermis, there is an extensive attenuation of collagen in the atrophic dermis and skin appendages are absent.^(2,3)

Treatment is indicated only for aesthetic concerns. Malignant changes and systematic abnormalities are usually rare.⁽³⁾ Recurrence is rare post-surgical excision, hence surgical excision is considered as the best choice of treatment.^(4,7)

We report a case of NLCS due to its rarity and its presentation in an unusual site that was clinically compatible with the classical type

concerning age and lesion's characteristics. The histopathological findings were typical and consistent with NLCP which finalizes the diagnosis.

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