ORIGINAL RESEARCH PAPER

Pathology

CASE REPORT :-NEVUS LIPOMATOSIS SUPERFICIALIS- AN UNUSUAL HAMARTOMA

KEY WORDS: Connective tissue, Hamartoma, Skin neoplasms

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ABSTRACT

Nevus lipomatosus cutaneous superficialis is a rare benign hamartomatous skin tumor characterized by dermal deposition of mature adipose tissue.Two clinical forms have been described (classical and solitary types).We describe a case of nevus lipomatosus cutaneous superficialis with a 23-year history of growth in a young male who had a solitary skin-colored tumoral mass on the right buttock. Histopathological findings were typical and confirmed the diagnosis. In this case, the lesion was a skin-colored isolated mass, as described in the solitary type, but its localization and age of appearance were compatible with the classical type.The combination of simultaneous clinical findings of both types had not been published before.

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) was first described in 1921 by Hoffman and Zurhelle.1 is a rare hamartoma of skin composed of admixture of mature adipose tissue, collagen bundles.2,3 It is usually sporadic and seen in childhood . There is no genetic or gender predilection.4,5 The commoner classical variant presents as multiple , skin-colored, pedunculated nodules. The uncommon solitary variant presents as a dome-shaped sessile papule or nodule.6

The pathogenesis of NLCS is not known. One theory suggests fat deposition to be a result of degenerative changes in connective tissues,.7 Other theories suggests that multiple type NLCS arises from adipose metaplasia in the course of degenerative changes in the dermal connective tissue. Other authors propose that the adipocytes represent a true nevus resulting from the developmental displacement of adipose tissue.8 The mature adipocytes may be 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 with a 23-year history of growth in a young male

CASE REPORT

A 23-year-old healthy man presented with an asymptomatic solitary mass over gluteal cleft, which has increased in size over the last 6-7 months.There was no family history of similar lesions.

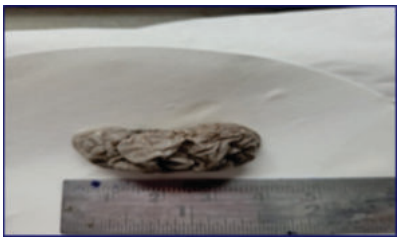

Figure 1

Physical examination revealed a skin-colored, pedunculated soft mass with a cerebriform surface and centrally located comedo-like plugs, measuring 12x8cm, on the right buttock (Figures 1 and and2).2). There was no ulceration, pigmentation, café-au-lait macules. Systemic examination was unremarkable

Skin-colored pedunculated mass with cerebriform surface on the right buttock

Figure 2

Detail of the lesion, showing some comedo-like plugs on its surface

An incisional skin biopsy was performed Histopatholoy analysis showed epidermis showing slight hyperkeratosis, acanthosis, and elongation of the rete ridges of the epidermis. Collections of mature adipose tissue were seen in the dermis interposed among the collagen bundles,There was no connection of the adipose tissue to the subcutaneous fat. We found focal spindle cell infiltrates surrounding the blood vessels (Figure 3). These findings supported the clinical diagnosis of NLCS.

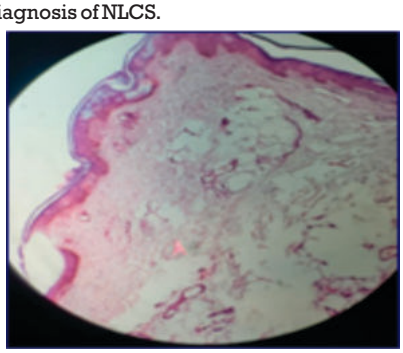

Figure 3

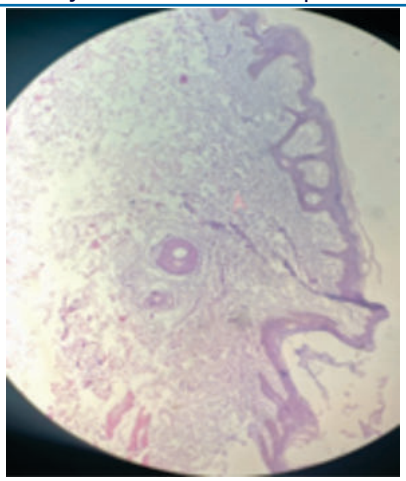

Figure 4

(Microphotograph showing adipose tissue interspersed among collagen bundles.)

Aggregates of ectopic adipocytes diffusely scattered between collagen fibers in the dermis, without connection with the subcutaneous tissue. (Hematoxylin & eosin X20)

The patient was submitted to a surgical excision with no recurrence at a six-month follow-up.

DISCUSSION

NLCS is a rare idiopathic hamartomatous anomaly that is characterized by ectopically situated mature adipocytes in the dermis.4,7 It may be present at birth or may even begin in infancy. It is classified into two clinical forms. The classical (or multiple) form occurs within the first three decades of life and is characterized by clusters of soft, yellow or skin-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 The solitary type can occur anywhere on the skin and usually appears during the third to sixth decades of life 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 In our case, although the patient had a solitary mass, the age of appearance, site of the lesion, and the dermatological characteristics were compatible with the classical form.

In both types, the lesions are asymptomatic, as seen in our patient. Ulceration occurs rarely, especially after external traumas or ischemia.2,7 Furthermore, café-au-lait macules, leukodermic spots, overlying hypertrichosis and comedo-like alterations can coexist.3,7 Similarly, in this report, the surface of the nevus was studded with some open comedones.

The histopathology of NLCS usually reveals a growth of mature adipocytes in the reticular dermis usually composing from 10-50% of the nevus.5,10 The adipocytes are usually seen forming collections around blood vessels or eccrine glands. There may be dense 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, showing more than 15% of the dermis replaced by adipose tissue, with focal spindle-shaped cell infiltration surrounding the blood vessels. We did not observe any changes like increased collagen density , decrease in elastic tissue, or adnexal fibrosis.

Clinically, the differential diagnosis includes nevus sebaceous, connective tissue nevus, neurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (Goltz syndrome).2,7 Histopathological studies usually sufficient to confirm diagnosis. Similar dermal collections of adipocytes are also present in some melanocytic nevi, pedunculated lipofibromas, 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 cosmetic reasons since malignant degeneration and systemic abnormalities are rare.3 Treatment is surgical excision. Wise excision is not required as recurrences' are rare.4,7

NLCS is a rare diagnosis. We suspect that many cases may be missed and reported as a hamartoma without the specific nomenclature. Early diagnosis is beneficial as they can grow quite large leading to aesthetic issues and /or discomfort. Our patient aand presentation as a solitary mass that was clinically compatible with the classical type. Typical histopathological findings helped in confirmation the diagnosis.

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