

BRIEF ARTICLE

Nevus Lipomatosus Cutaneous Superficialis: A Case Report in Egypt

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ABSTRACT

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare hamartomatous condition characterized by ectopic mature adipose tissue. We are reporting a case of a 12-year-old female with soft, non-tender, pedunculated nodules with a cerebriform surface over her right lower back. The lesion fulfilled the criteria of the classic type of NLCS and was surgically removed with no visible recurrence on follow up.

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a type of skin hamartoma characterized by the presence of ectopic mature adipose tissue in the dermis. This condition was first described in 1921 by Erich Hoffmann and Emil Zurhelle.¹ Classically, it presents as multiple groups of soft, pedunculated, yellow or skin-colored nodules, papules, or plaques in the pelvic girdle region.² This presentation is known as the classic type, or Nevus of Hoffmann-Zurhelle. It may also present as a dome-shaped sessile papule, known as the solitary type.³ Different theories have been proposed describing the pathogenesis of this condition, however, the true underlying mechanism is still unclear.⁴

A 12-year-old female with Fitzpatrick skin type III presented to the outpatient clinic with a solitary mass on her right lower back. The lesion was first noted by the patient's mother around the age of 9.5 years and continued to gradually increase in size over the next 2.5 years. The main complaint was limited to the unpleasant shape with no history of similar lesions reported in other family members.

Physical examination revealed a skin-colored, pedunculated, cauliflower-like mass with a cerebriform surface, measuring 5×5 cm, on the right lower back. (**Figure 1**) There was no associated pigmentation, hair growth, or ulcerations. Systemic examination was unremarkable except for sensorineural hearing loss, which was also reported in the patient's father and paternal uncles.

An excision biopsy with safety margins was performed. Histopathological analysis showed replacement of the dermal collagen

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by mature adipocytes, not connected to the subcutaneous fat, with no nuclear atypia or mitotic figure, which supports the diagnosis of NLCS. (Figure 2). The lesion was surgically removed with no recurrence detected on follow up at 6 months.

DISCUSSION

NLCS is a rare hamartomatous skin lesion that has been reported in different locations around the world.^{4, 5} Two main forms of this condition have been identified. The classic form presents as multiple lesions within the first three decades of life or at birth. Grossly, it presents as clusters of soft, yellow or skin-colored nodules, papules, or plaques located at the lower trunk, especially in the pelvic girdle and gluteal, sacral, or lumbar regions.¹ The solitary type presents later in life, between the third and sixth decades, at any location of the body, including the mucus membranes.⁶ A single lesion was detected in our case, however, it has the characteristics of the classic type. Although several concomitant features were reported as an association with the primary lesion, such as comedo-like plug, overgrowth into giant NLCS, and ulcerations, none of these features were observed in the present case.^{7, 8}

Several hypotheses regarding the pathogenesis of this lesion are proposed in the literature. Some of these hypotheses include this lesion being a true nevus as a result of developmental displacement of adipose tissue or adipose metaplasia. Another theory describes this lesion as differentiating lipoblasts in perivascular areas that developed into mononuclear cells and subsequently grew into mature adipocytes.^{4, 8} However, the exact underlying mechanism of this lesion is still unclear.

In this case, the patient suffered from sensorineural hearing loss with a positive family history in the father and paternal uncles. While this could be a coincidental finding, it raises the suspicion for a possible underlying mechanism at the genetic level that may explain this association, providing that chromosomal deletions have been reported in several NLCS cases.^{9, 10} NLCS has been reported to recur following resection⁴, but the recurrence rate is generally very low. In the present case, no recurrence was observed on follow up at 6 months.

CONCLUSION

It is important to know the characteristics of the present condition, as it may be confused with other skin disorders such as skin tags and nevus sebaceous. Thus, knowing all the variable presentations of this lesion may help to fulfill this need. Further studies are needed to investigate the possible role of chromosomal abnormalities in the development of NLCS.

Informed consent: Written informed consent was obtained from the patient's parents for publication of the details of their medical case and any accompanying images.

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Figure 1. Skin-colored, pedunculated, cauliflower-like mass with a cerebriform surface, measuring 5×5 cm, on the right lower back.

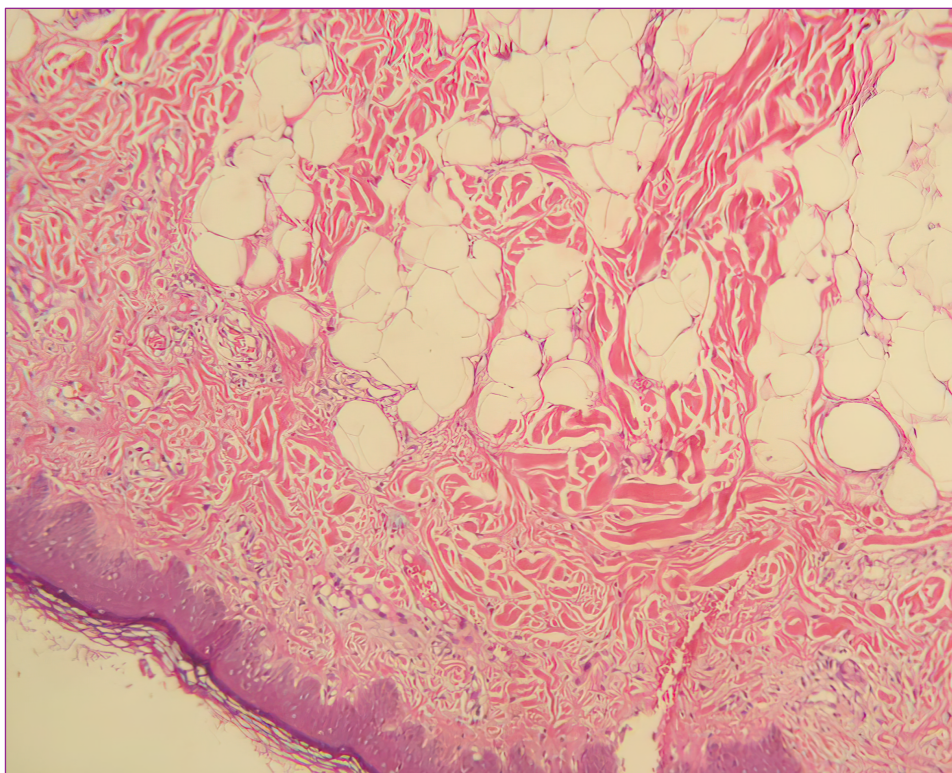


Figure 2. Nevus lipomatosus cutaneus superficialis covered by non-ulcerated skin epidermis that shows dermal collagen replaced by mature adipocytes.