Iran Red Crescent Med J. 2020 June; 22(6):e99710. doi: 10.5812/ircmj.99710.

Published online 2020 July 8. **Case Report**

Nevus Lipomatosus Cutaneous Superficialis: A Case Report and Literature Review

Raymond Dieu Kiat Yeak [1,*], Yee Yee Yap [2] and Nizlan M. Nasir [1]

[1]Department of Orthopaedic Surgery, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang, Malaysia
[2]Department of Haematology, Ampang Hospital, Ampang, Malaysia

[*]*Corresponding author*: Department of Orthopaedic Surgery, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang, Malaysia. Tel: +60-162155603, Email: rayyeak@yahoo.com

Received 2019 November 26; **Revised** 2020 February 26; **Accepted** 2020 March 18.

Abstract

Introduction: Nevus lipomatosus cutaneous superficialis (NLCS) is uncommon and can be encountered in various medical specialties. In this study, we report a rare case of solitary pedunculated nevus lipomatosus cutaneous superficialis in a middle-aged female patient and highlight its clinical and histopathological features.
Case Presentation: A 43-year-old Malaysian female patient presented to our center with a 2-cm long and 1-cm wide solitary, flesh-colored nodule over the anterolateral aspect of her thigh. Excisional biopsy was performed, and the histopathology report showed NLCS. There are three types of NLCS, namely the solitary, multiple, and generalized forms. The etiology of NLCS is usually idiopathic, with no gender or familial preponderance. NLCS is usually misdiagnosed and should be differentiated from nevus sebaceous, connective tissue nevus, neurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (also known as Goltz syndrome).
Conclusions: In conclusion, NLCS is rare and benign condition, which should be differentiated from other malignant skin conditions as it is commonly misdiagnosed. Therefore, physicians and surgeons should be aware of its clinical and histopathological features.

Keywords: Connective Tissue, Hamartoma, Skin Neoplasms

1. Introduction

Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon condition with classical and solitary types. NLCS was first reported by Hoffman and Zurhelle in 1921 (1). In this study, we report a rare case of solitary pedunculated NLCS in a middle-aged female patient and highlight its clinical and histopathological features.

2. Case Presentation

A 43-year-old Malaysian female patient presented to a private specialist hospital with a 2-cm long and 1-cm wide swelling over the anterolateral aspect of her thigh (Table 1). It was measured using a 6-inch disposable paper ruler (Aspen Surgical, Caledonia, MI, USA), calibrated in inches and centimeters. It was painless and had been growing in size for the past two years from September 2017 till September 2019. Physical examination revealed a solitary, flesh-colored nodule with no café au lait macules or skin lesions elsewhere (Figure 1). Excisional biopsy was performed, and the histopathological report demonstrated a papillomatous lesion covered by squamous epithelium with lobules of adipose tissue interspersed among the collagen bundles in the stroma (Figure 2B-D). The adnexal glands were entrapped within the fat lobules (Figure 2A). The epidermis showed mild acanthosis and hyperkeratosis (Figure 2B-D). The patient was then diagnosed with NLCS.

3. Discussion

Hoffman and Zurhelle described the classical NLCS as multiple, non-tender, soft, pedunculated, cerebriform, yellowish, or skin-colored papules or nodules. 1 The classical type usually presents in the first two to three decades of life (1).

Solitary NLCS, on the other hand, is rarer and can present as a solitary dome-shaped sessile papule or nodule (2). It usually occurs during the third to sixth decades of life and can occur anywhere in the body (3).

There is another rarely described type, which is the generalized form (4). This type was first described in 1969

Table 1. Summary of the Clinical, Nature, Macroscopic and Microscopic Appearance of the Lesion

Clinical	Nature	Macroscopic Appearance	Microscopic Appearance
2 cm long × 1 cm wide solitary swelling	A solitary, pedunculated, flesh-colored nodule	Skin ellipse with underlying subcutaneous tissue with a central greyish nodule	A papillomatous lesion covered by squamous epithelium with lobules of adipose tissue interspersed among the collagen bundles in the stroma. The adnexal glands were entrapped within the fat lobules. The epidermis showed mild acanthosis and hyperkeratosis

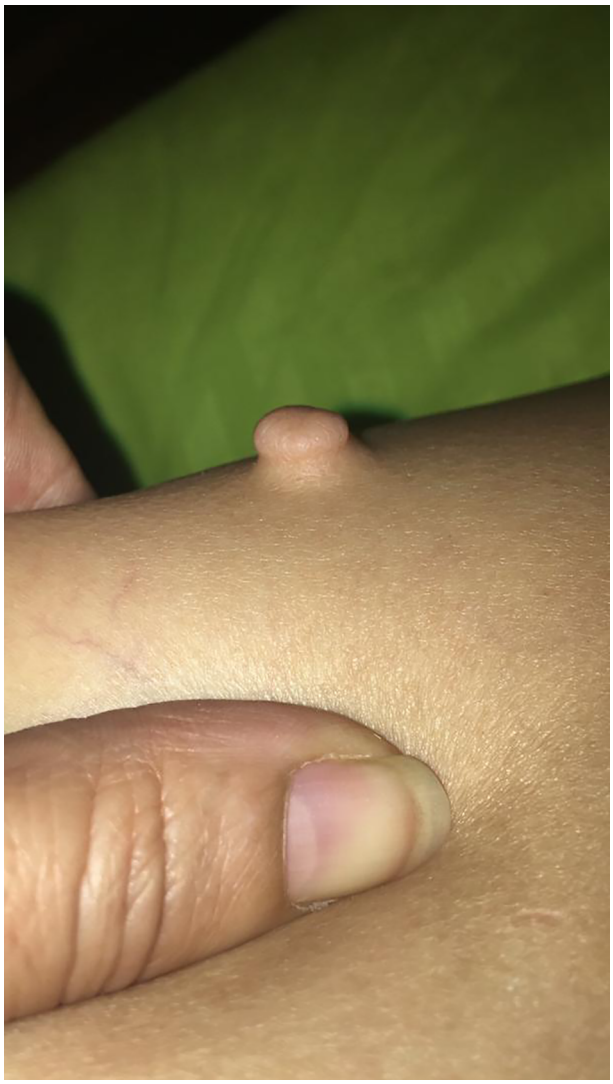

Figure 1. The anterolateral aspect of the patient's thigh showing a solitary, flesh-colored nodule

by Ross and is also known as underlying nevus lipomatosis (Table 2) (4). It is characterized by an excess of folded skin called the Michelin tire baby syndrome (MTBS) (4). It may be deep and extensive and linked to systemic manifestations. Bass et al. (5) suggested an autosomal mode of inheritance. Genetic defect has also been suggested to be the cause of the disease. The UNL has been linked to chromosome 11 deletion (6). Another study has linked the disease to the genetic defect of 2p24 deletion (7). Goucha et al. (2) found that the solitary type is rarer in comparison to the multiple type. The most common sites are the pelvic girdle, the lower trunk, the gluteal region, and the thigh (2). However, another study showed that the multiple type was more common among the patients (8). There is no gender or familial preponderance in this disease (9).

Table 2. Comparison of Previous Case Reports

Author	Case Reports
Ross et al. (4)	The first case of marked folding of the skin with underlying nevus lipomatosus, which occurred in his patient in South Africa.
Gardner et al. (6)	The second reported case of marked folding of the skin, with an underlying nevus lipomatosus.
Cardot-Leccia et al. (7)	Described the first cytogenetic aberration in a case of localized NLCS of Hoffmann and Zurhelle (HZ) associated with a loss of chromosome 2p24-pter.
Chopra et al. (10)	A 6-year-old boy with a gluteal swelling with 2 unusual histological findings.
Dhamija et al. (11)	Reported a case of adult-onset classical NLCS on perianal area.
Takegawa et al. (12)	Reported a case of giant NLCS on the buttock.

The etiology of NLCS is usually idiopathic (10). However, it has been postulated that changes in dermal collagen and elastic tissues lead to fat cell depositions among collagen bundles in the superficial dermis (13). It has also been proposed that the precursor mesenchymal perivascular cells develop to adipocytes in the dermis (13).

NLCS is usually misdiagnosed and should be differentiated from nevus sebaceous, connective tissue nevus, neurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (also known as Goltz syndrome) (11). The disease is commonly diagnosed through histopathology, but the pathological tissue can be confused with a plexiform neurofibroma, connective tissue nevus, vascular malformation, lipomatosis, or lipoblastomatosis (12). The preferred treatment is excision usually performed for cosmetic reasons. Other reported treatments include car-

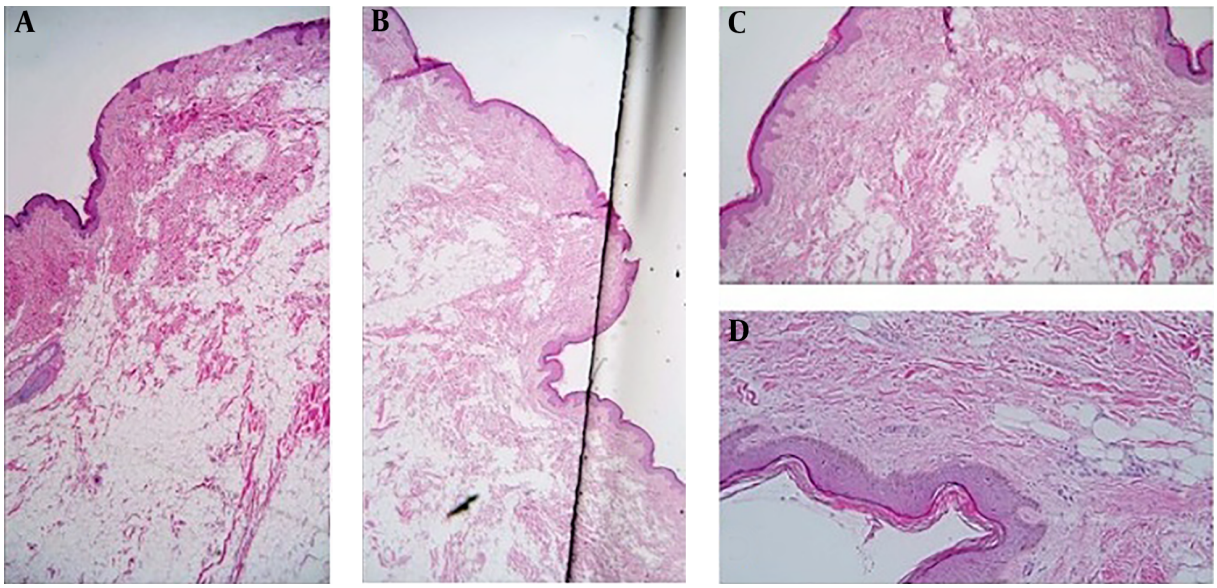

Figure 2. A, The adnexal glands entrapped within the fat lobules (H & E; 4×); B-D, the histopathological report demonstrated a papillomatous lesion covered by squamous epithelium with lobules of adipose tissue interspersed among the collagen bundles in the stroma. The epidermis shows mild acanthosis and hyperkeratosis (B: H & E; 4×; C: 10×; D: 20×).

bon dioxide laser treatment, intralesional phosphatidylcholine, and sodium deoxycholate injection, which have shown promising results (14, 15).

3.1. Conclusions

NLCS is a rare and benign condition, which should be differentiated from malignant skin conditions as it is commonly misdiagnosed. Therefore, physicians and surgeons should be aware of its clinical and histopathological features.

Footnotes

Authors' Contribution: RDKY developed the original idea, abstracted and wrote the manuscript. YYY and NMN contributed to the development of the abstract and helped to prepare the manuscript.

Conflict of Interests: None to declare.

Ethical Approval: This is a case report and does not require ethical approval.

Funding/Support: None.

Informed Consent: Informed consent was obtained.

References

1. Hoffmann E, Zurhelle E. Über einen Naevus lipomatodes cutaneus superficialis der linken Glutaalgegend. *Archiv fur Dermatologie und Syphilis*. 1921;**130**(1):327–33. doi: 10.1007/bf01826150.
2. Goucha S, Khaled A, Zeglaoui F, Rammeh S, Zermani R, Fazaa B. Nevus lipomatosus cutaneous superficialis: Report of eight cases. *Dermatol Ther (Heidelb)*. 2011;**1**(2):25–30. doi: 10.1007/s13555-011-0006-y. [PubMed: 22984661]. [PubMed Central: PMC3437641].
3. Ranjkesh MR, Herizchi QH, Yousefi N. Nevus lipomatosus cutaneous superficialis: A case report with histologic findings. *J Turk Acad Dermatol*. 2009;**3**(1):93103c.
4. Ross CM. Generalized folded skin with underlying lipomatous nevus: the Michelin Tyre baby. *Arch Dermatol*. 1972;**106**(5):766. [PubMed: 4635809].
5. Bass HN, Caldwell S, Brooks BS. Michelin tire baby syndrome: familial constriction bands during infancy and early childhood in four generations. *Am J Med Genet*. 1993;**45**(3):370–2. doi: 10.1002/ajmg.1320450318. [PubMed: 8434626].
6. Gardner EW, Miller HM, Lowney ED. Folded skin associated with underlying nevus lipomatosus. *Arch Dermatol*. 1979;**115**(8):978–9. [PubMed: 464627].
7. Cardot-Leccia N, Italiano A, Monteil MC, Basc E, Perrin C, Pedeutour F. Naevus lipomatosus superficialis: A case report with a 2p24 deletion. *Br J Dermatol*. 2007;**156**(2):380–1. doi: 10.1111/j.1365-2133.2006.07622.x. [PubMed: 17223884].
8. Triki S, Mekni A, Haouet S, Mokni M, Kchir N, Ben Osman Dhahri A, et al. [Nevus lipomatosus cutaneous superficialis: A clinico-pathological study of 13 cases]. *Tunis Med*. 2006;**84**(12):800–2. French. [PubMed: 17288284].
9. Dotz W, Prioleau PG. Nevus lipomatosus cutaneus superficialis. A light and electron microscopic study. *Arch Dermatol*. 1984;**120**(3):376–9. [PubMed: 6231000].
10. Chopra R, Al Marzooq YM, Siddiqui FA, Aldawsari S, Al Ameer A. Nevus lipomatosus cutaneous superficialis with focal lipocytic pagetoid epidermal spread and secondary calcinosis cutis: A case report. *Am J Dermatopathol*. 2015;**37**(4):326–8. doi: 10.1097/DAD.0000000000000118. [PubMed: 25014105].

11. Dhamija A, Meherda A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: An unusual presentation. *Indian Dermatol Online J*. 2012;**3**(3):196–8. doi: 10.4103/2229-5178.101819. [PubMed: 23189254]. [PubMed Central: PMC3505429].
12. Takegawa M, Kakudo N, Morimoto N, Hihara M, Masuoka H, Kusumoto K. Giant nevus lipomatosus cutaneous superficialis on the buttock. *Plast Reconstr Surg Glob Open*. 2018;**6**(11). e1918. doi: 10.1097/GOX.0000000000001918. [PubMed: 30881777]. [PubMed Central: PMC6414102].
13. Holtz KH. [Histology of naevus lipomatodes cutaneous superficialis (Hoffmann-Zurelle)]. *Arch Klin Exp Dermatol*. 1955;**199**(3):275–86. German. [PubMed: 14388675].
14. Sardana K, Bansal S, Garg VK, Khurana N. Treatment of Nevus lipomatosus cutaneous superficialis with CO2 laser. *J Cosmet Dermatol*. 2017;**16**(3):333–5. doi: 10.1111/jocd.12327. [PubMed: 28317240].
15. Kim HS, Park YM, Kim HO, Lee JY. Intralesional phosphatidylcholine and sodium deoxycholate: A possible treatment option for nevus lipomatosus superficialis. *Pediatr Dermatol*. 2012;**29**(1):119–21. doi: 10.1111/j.1525-1470.2011.01413.x. [PubMed: 22150229].