

Nevus Lipomatosus Cutaneous Superficialis - A Rare Hamartoma

Suchibrata Das,¹ MD, Joyeeta Chowdhury,¹ MD, Subhamay Neogi,¹ MD, Sangita Patra,² MD

Address: ¹Department of Dermatology , Venereology and Leprosy. N R S Medical College, 138 A J A Bose Road, Kolkata 700014, West Bengal , India, ²Department of Gynaecology and Obstetrics, Kolkata Medical College and Hospital , Kolkata , 88 College Street, Kolkata.

E-mail: suchibratadas@yahoo.com

* Corresponding Author: Dr. Suchibrata Das, Department of Dermatology , Venereology and Leprosy. N R S Medical College, 138 A J A Bose Road, Kolkata, West Bengal, India.

Published:

J Turk Acad Dermatol 2016; 10 (3): 16103c3

This article is available from: <http://www.jtad.org/2016/3/jtad16103c3.pdf>

Keywords: Nevus lipomatosus superficialis, comedones

Abstract

Observation: Nevus lipomatosus cutaneous superficialis of Hoffmann-Zurhelle is a rare cutaneous hamartoma of adipocytes. Classical and solitary types are found. Histological findings are groups and strands of fat cells, found embedded among the collagen bundle of the dermis, often as high as the papillary dermis. Our patient is a case of classical variant of Nevus lipomatosus cutaneous superficialis present on left gluteal region with multiple cerebriform nodules with comedone like plugs on its surface.

Introduction

Nevus lipomatosus cutaneous superficialis (NLCS) of Hoffmann-Zurhelle is a rare cutaneous hamartoma of adipocytes [1]. Classical and solitary types are found. Histological findings are groups and strands of fat cells, found embedded among the collagen bundle of the dermis, often as high as the papillary dermis [2]. We present a case of classical variant of Nevus lipomatosus cutaneous superficialis (NLCS).

Case Report

A 28 year old female patient presented to the dermatology OPD with multiple asymptomatic growths over upper outer quadrant of left buttock since last 8 years. There were multiple asymptomatic, yellowish to skin coloured, smooth surfaced, soft nodules coalescing to form two cerebriform plaques measuring 5x4 cm and 4 cm

circular (Figure 1). There was also a single soft nodule in the vicinity of the lesion. Many comedone like plugs were present on the surface. There was no tenderness, ulceration or induration nor any regional lymphadenopathy. Rest of the mucocutaneous, systemic and laboratory findings were normal. Histopathological examination of the incisional biopsy specimen showed aggregation of mature adipocytes in dermis (as high as in the papillary dermis) interposed among collagen bundles occupying more than half of dermis (Figure 2) with no connection of these adipocytes with the subcutaneous fat. The adipose tissues were not encapsulated. A diagnosis of Nevus lipomatosus cutaneous superficialis (NLCS) was done based on the histologic findings.

Discussion

NLCS is an uncommon, idiopathic hamartomatous benign condition characterized by the



Figure 1. Skin coloured nodules with cerebriform surface, comedo like plugs seen on left gluteal region

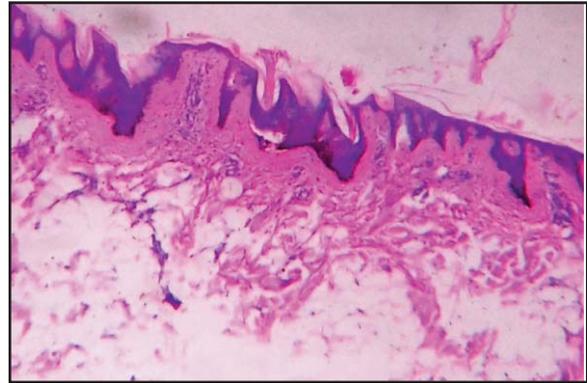


Figure 2. Lobule of mature adipose tissue separated by bundles of collagen extending in the reticular dermis. (H & E X40)

presence of ectopic mature adipose tissue within the dermis. It is classified into two clinical variants: the classical form and solitary form. The classical form usually composed of multiple and grouped skin-colored, pedunculated and cerebriform nodules that often coalesce to form a plaque, with zosteriform, linear or segmental distribution. These are usually seen during the first two decades of life, after which they become infrequent [3,4]. It is slow-growing, with a smooth or cerebriform surface, and can reach a large size if left untreated. The largest size reported so far has been 40 cm x 28 cm [5]. The most common sites are the pelvic girdle, the lower trunk, the gluteal region, and the thigh [6,7]. It may be present at birth or may even begin in infancy (nevus angioliipomatosus of Howell). If the disorder begins during infancy, the change of hypoplastic dermis leads to pseudotumor yellow protrusions concurrent with skeletal and other malformations [3,4]. The second clinical pattern of NLCS is a solitary papule or nodule mimicking skin tag [7]. It usually appears later than the classical form, in third to sixth decades of life [6,8]. Any site can be involved, including unusual ones like the scalp, the eyelid, the nose, and the clitoris [9].

NLCS is usually asymptomatic as noted in our case. Rarely, ulceration may occur, due to external trauma or ischemia [9]. Café-au-lait macules, leukodermic spots, overlying hypertrichosis and comedo-like alterations sometimes coexist [4,7,10]. In our case only comedo like lesions were present on the surface.

Histopathologically, epidermal changes are not always present but “mild to moderate acanthosis, basket weave hyperkeratosis, increased basal pigmentation and focal elongation of rete ridges” have been noted [8]. Dermal collagen bundles show fat cells that have frequent extension to the papillary layer. In instances with relatively large amounts of fat, fat lobules are irregularly distributed throughout the dermis when the amount of fat is relatively large and the boundary between the dermis and hypodermis is ill-defined or lost. The fat may all be mature, but in some instances an occasional small, incompletely lipidized cell may be observed. Usually the dermis is normal even with the fat cells but in some cases of NLCS there is an increase in the density of collagen bundles, fibroblasts and blood vessels in the dermis [4,10].

The pathogenesis of NLCS remains unknown and several theories have been proposed: *Hoffman and Zuhrelle* postulated that fat deposition in the dermis is secondary to degenerative changes (metaplasia) in the connective tissue [2]. Other authors hypothesized that adipocytes originate from the pericytes of dermal vessels [11]. For others, fat cells represented a true nevus that resulted from the focal heterotopic development of adipose tissue [12]. Electron microscopic findings strongly confirmed the perivascular origin of young adipocytes and the differentiation into mature fat [11]. Recently, a report of a NLCS with a 2p24 deletion has been published. Nevus Lipomatosis Cutaneous Superficialis should be differentiated from skin tags,

neurofibroma, lymphangeoma, haeman-geoma and focal dermal hypoplasia (Goltz Syndrome). Histopathological evaluation usually helps in differentiation.

Treatment is not necessary other than for cosmetic reasons. Systemic abnormalities and malignant changes have not been associated with NLCS. Excision is curative and recurrence after surgery is rare.

References

1. Bancalari E, Martínez-Sánchez D, Tardío JC. Nevus lipomatosus superficialis with a folliculosebaceous component: report of 2 cases. *Patholog Res Int* 2011; 18: 105973. PMID: 21559190
2. Ragsdale BD. Tumours With Fatty, Muscular, Osseous, and Cartilaginous Differentiation In: *Lever's Histopathology of The Skin*. Elder DE, Elenitsas R, Jhonson BL, Murphy GF, editors. 9th ed. Philadelphia: Lippincott Williams & Wilkins, 2005, 1061-1107.
3. Chougule A, Kumari R, Thappa DM. Giant nevus lipomatosus cutaneous superficialis of the thigh. *Indian J Dermatol* 2007; 52: 120-121.
4. Das JK, Sengupta S, Gangopadhyay AK. Nevus lipomatosus superficialis over neck, an atypical site. *Indian J Dermatol Venereol Leprol* 2006; 72: 66-67. PMID: 16481721
5. Yap FBB. Nevus lipomatosus superficialis. *Singapore Med J* 2009; 50: 161-162. PMID: 19495497
6. Ioannidou DJ, Stefanidou MP, Panayiotides JG, Tosca AD. Nevus lipomatosus cutaneous superficialis (Hoffmann-Zuhrelle) with localized scleroderma like appearance. *Int J Dermatol* 2001; 40: 54-57. PMID: 11277956
7. Khandpur S, Nagpal SA, Chandra S, Sharma VK, Kaushal S, Safaya R. Giant nevus lipomatosus cutaneous superficialis. *Indian J Dermatol Venereol Leprol* 2009; 75: 407-408. PMID: 19584473
8. Buch Archana C, Paniker NK, Karve PP. Solitary nevus lipomatosus cutaneous superficialis. *J Postgrad Med* 2005; 51: 47-48. PMID: 15793341
9. Goucha S, Khaled A, Zéglauoui F, Rammeh S, Zermani R, Fazaa B. Nevus lipomatosus cutaneous superficialis: Report of eight cases. *Dermatol Ther (Heidelb)* 2011; 1: 25-30. PMID: 22984661
10. Dhamija A, Meherda A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: An unusual presentation. *Indian Dermatol Online J* 2012; 3: 196-198. PMID: 23189254
11. Lane JE, Clark E, Marzec T. Nevus lipomatosus cutaneous superficialis. *Pediatr Dermatol* 2003; 20: 313-314. PMID: 12869151
12. Dotz W, Prioleau PG. Nevus lipomatosus cutaneous superficialis. *Arch Dermatol* 1984; 120: 376-379. PMID: 6231000