# Nevus Lipomatosus Cutaneous Superficialis of Hoffmann and Zurhelle: a rare cutaneous hamartoma

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## ABSTRACT

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare idiopathic cutaneous hamartoma characterized by ectopic clusters of mature adipose tissues in dermis. It is classified into two types. Classical type presenting as multiple lesions with onset at birth or within third decade of life and solitary type with onset between third to sixth decade of life. Here we present a case of 45 years female with adult onset, asymptomatic, grouped, skin colored, soft sessile growth in zosteriform pattern on left buttock. We here intend to report rare case of classical Nevus Lipomatosus Cutaneous Superficialis.

## KEY WORDS

*Hoffmann, Nevus lipomatosus cutaneous superficialis, Zurhelle*

## INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare cutaneous hamartoma characterized by aggregates of mature ectopic adiopocytes in the dermis.[1] It was first described in 1921 by Hoffman and Zurhelle.[2] Two clinical types are recognized: classical and solitary type. The classical type is rare, usually unilateral, band-like, linear, or zosteriform in distribution.[1] It is characterized by groups of multiple, non-tender, soft, pedunculated or sessile, cerebriform, yellowish or skin-colored papules, nodules or plaques.[1,3] The solitary type is more common, characterized by dome shaped or sessile papule, resembling the skin tag. We present a rare case of classical NLCS.

## CASE REPORT

A 45 years female presented with multiple, painless, skin lesions confined to the right gluteal region since 20 years. The lesion began as a single nodule and over time similar lesion appeared and became confluent to form linear plaque. Physical examination revealed multiple, skin colored, soft, pedunculated, cerebriform, non tender nodules and plaques that occupied an area of approximately 10×5 cm on the right gluteal region. The surface was studded with comedo like plug without ulceration or discharge. There was no hair growth and café au lait macules. Systemic examination was unremarkable.

Skin biopsy was performed and microscopy showed nests of ectopic mature adipocytes in dermis. The diagnosis of NLCS was made based on clinical examination and biopsy findings.

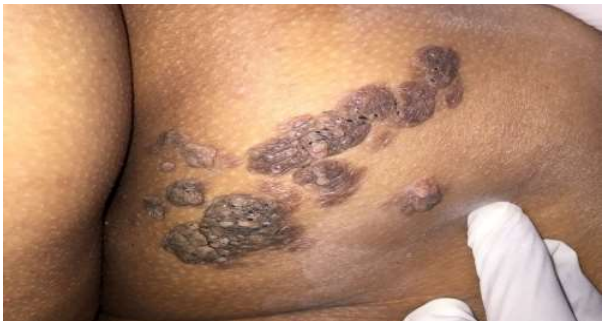

**Figure 1. Nevus lipomatosus on right gluteal area showing soft, grouped, sessile and pedunculated papules and plaques with central comedo like plug**

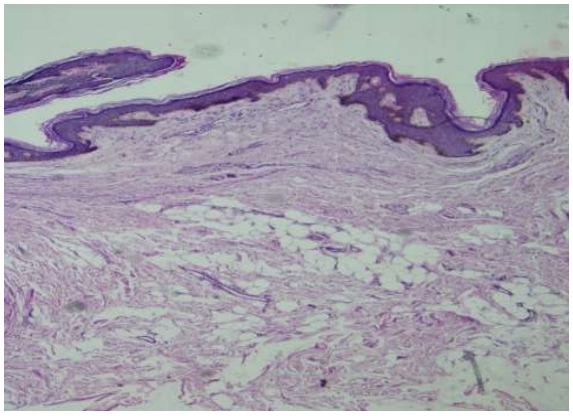

**Figure 2. H&E stain (10×) showing nests of mature adipocytes encroaching the dermis**

## DISCUSSION

Nevus lipomatosus cutaneous superficialis (NLCS) is a developmental anomaly of unknown origin characterized by the presence of ectopic mature adipocytes in the papillary and reticular dermis.[4,5]

The classical type was first recognized by Hoffman and Zurhelle in 1921 and may be present at birth, infancy (nevus angiolipomatosus of Howell) or during the first three decade of life.[4] It is characterized by multiple, soft, cerebriform, sessile or pedunculated papules, nodules and plaque arranged in zones or zosteriform pattern.[4] These slow growing tumors are most commonly located at lower back, pelvic girdle, buttocks and upper thigh.

The second clinical type is a solitary papule or nodule resembling skin tag with the onset during third to six decade of life. This clinical form can occur anywhere on the skin. It has been reported in rare sites such as face scalp eyelids and clitoris.[4,5] No family or sex predilection has been reported.[6]

The lesion is usually asymptomatic as seen in our patient. Occasionally they may ulcerate secondary to external trauma or ischemia.[1] There are reports of coexistent café au lait macules, leukoderma, hypertrichosis and comedo like opening with NCLS. Several authors have reported as association of follicular papules, hypertrophic pilosebaceous unit, angiokeratoma of Fordyce and hemangioma with NCLS.[7] Conversion to malignancy has not been reported.

The exact pathogenesis is unknown, however many theories has been put forward. Hoffman and Zurhelle proposed that fat deposition in the dermis is secondary to degenerative changes (metaplasia) in the connective tissue.[7] Other theories include developmental displacement (heterotopia) of adipocytes and mesenchymal perivascular cells (primitive lipoblast) subsequently transform into mature adipocytes.[6] The role of genetic abnormality is still unknown, but a case report of NLCS showing 2p24 deletion has been published.[8]

Histopathology shows ectopic mature adipocytes, intermingled with collagen bundles in the dermis and the perivascular area.[7] The adipocytes may have connection to underlying subcutaneous fat or be separated by collagen.[1] Perivascular infiltration of dermis and subcutis with chronic inflammatory cells.[6]

NCLS has to be differentiated clinically from nevus sebaceous, neurofibroma, lymphangioma, focal dermal hypoplasia, cylindroma, trichoepithelioma, and angiolipoma.[7]

Treatment is needed for only cosmetic reason. Surgical excision is the treatment of choice and post-surgical recurrence is rare.[6,7] Cryotherapy yields partial though satisfactory results.[6,7] Surgical excision is useful for solitary NCLS, but impractical for classic type due to its larger size. $CO_2$ laser has been reported to be highly effective, well-tolerated and less invasive.[9]

## REFERENCES


1. Dhamija A, Meherda A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: an unusual presentation. *Indian Dermatol Online J*. 2012;3:196-8.
2. Dotz W, Prioleau PG. Nevus lipomatosus cutaneous superficialis. *Arch Dermatol*. 1984;120:376-9.
3. Yap FB. Nevus lipomatosus superficialis. *Singapore Med J*. 2009;50:e161–2.
4. Patil SB, Narchal S, Paricharak M, More S. Nevus lipomatosus cutaneous superficialis: a rare case report. *Iran J Med Sci*. 2014;39:304–7.
5. Lima CD, Lemes LR, Melo DF. Yellow dots in trichoscopy: relevance, clinical significance and peculiarities. *Anais Brasileiros de Dermatologia.* 2017 Sep-Oct; 92(5): 711–13.
6. Khandpur S, Nagpal SA, Chandra S, Sharma VK, Kaushal S, Safaya R. Giant nevus lipomatosus cutaneous superficialis. *Indian J Dermatol Venereol Leprol*. 2009;75:407-8.
7. Goucha S, Khaled A, Zéglaoui F, Rammeh S, Zermani R, Fazaa B. Nevus lipomatosus cutaneous superficialis: Report of eight cases. *Dermatol Ther (Heidelb).* 2011 Dec;1(2): 25–30.
8. Cardot-Leccia N, Italiano A, Monteil MC, Basc E, Perrin C, Pedeutour F. Nevus lipomatosus superficialis: a case report with a 2p24 deletion. *Br J Dermatol*. 2007;156:380–81.
9. Fatah S, Ellis R, Seukeran DC, Carmichael AJ. Successful CO2 laser treatment of naevus lipomatosus cutaneous superficialis. Clin Exper Dermatol. 2010;35:559–60.