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Case Report

Naevus lipomatosus cutaneous superficialis: report of two cases

皮膚淺表脂肪性痣的兩例報告

V Kuntoji, C Kudligi, PV Bhagwat, R Rathod, S Krishna, S Vanjeri

Naevus lipomatosus cutaneous superficialis (NLCS) is an uncommon idiopathic hamartomatous condition characterised by groups of mature fat cells situated within the dermis. There are two types: the classical type as first described by Hoffmann and Zurhelle consists of multiple, soft, non-tender skin coloured to yellow papules and nodules which often coalesce to form plaques. The solitary type consists of solitary dome-shaped papule or nodule. We report two cases of classical type of NLCS which had varying clinical features and characteristic histopathological features.

皮膚淺表脂肪性痣是一種罕見的特發性錯構瘤病症，其特徵是眞皮內可見成熟脂肪細胞群。有兩種類型：霍夫曼和楚赫勒首先描述的經典型是多個柔軟無觸痛的皮膚色或黃色的丘疹或結節，常聚結形成斑塊；單生型則是單個拱頂形的丘疹或結節。我們報告了兩例具有不同臨床特徵和獨特組織病理學特徵的經典型皮膚淺表脂肪性痣。

Keywords: Adipocytes, dermis, hamartoma, naevus

關鍵詞：脂肪細胞、眞皮、錯構瘤、痣

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Introduction

Naevus lipomatosus cutaneous superficialis (NLCS) is a benign cutaneous hamartomatous condition characterised clinically by single or multiple soft nodules or papules and histologically by ectopic adipose tissue in the dermis.[1,2] The two clinical types recognised are the classical (known as Hoffman Zurhelle) and the solitary (known as pedunculated lipofibroma).[2] The clinical presentation of the former is seen as groups of yellowish to skin coloured multiple soft papules, nodules or plaques usually in the pelvic or gluteal

region and that of latter is seen as solitary papule or nodule with wider distribution in the skin like the ear, scalp, forehead, back, axilla, arm, upper thigh and knee.[2,3] We report two cases of NLCS both of which had varying clinical features of classical type of NLCS along with characteristic features on histology.

Case report

Case 1

A 10-year-old boy presented to our Department of Dermatology with asymptomatic nodules on the left buttock since birth which had gradually progressed in size and number to form a mass. There were no systemic complaints and family history was insignificant. On cutaneous examination, multiple skin coloured to yellowish nodules coalescing to form cerebriform mass of size 8x4 cm was seen over the left buttock. Satellite lesions of varying sizes were present 2 cm from the mass (Figure 1). Systemic examination was normal. Routine blood investigations were within normal limits. Histopathological examination of the biopsy specimen from cerebriform mass revealed mature lipocytes and fat interspersed within connective tissue which were non-encapsulated (Figure 2). The features were consistent with NLCS.

Case 2

A 30-year-old male presented to us with an asymptomatic raised skin lesion over his right buttock which was a small papule at birth and was gradually progressive. It was associated with occasional itching. On cutaneous examination, a solitary cerebriform skin coloured plaque of size 3x4 cm was present over right buttock (Figure 3). Multiple black coloured comedo-like lesions were present over the plaque. Routine blood investigations were normal. Histopathological examination revealed hyperkeratosis and increased melanin at basal layer. Papillary and reticular dermis showed lobules of adipose tissue

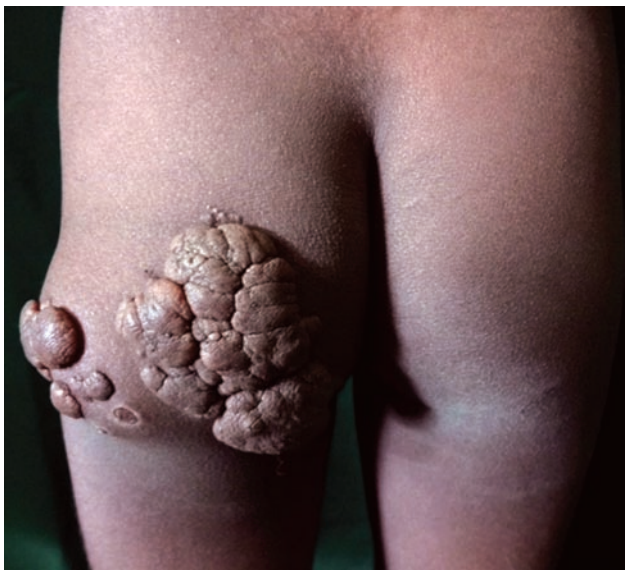

Figure 1. Cerebriform mass and satellite lesions over left buttock.

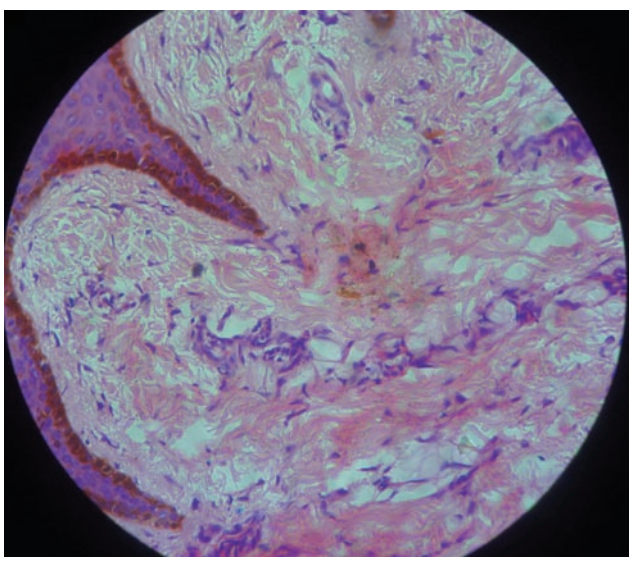

Figure 2. Mature lipocytes and fat interspersed within connective tissue (H and E: x40).

amidst hyalinised stroma, suggestive of NLCS (Figure 4).

Discussion

Naevus lipomatosus cutaneous superficialis is an uncommon, idiopathic hamartomatous condition characterised by dermal deposition of mature ectopic adipocytes. The proportion of fatty tissue varies from more than 50% to less than 10% of the dermis.[4] Hoffmann and Zurhelle in 1921 described the first of case of NLCS in a 25-year-old man who presented with multiple soft nodules in gluteal region. This was the classical form of NLCS. In 1968, Weitzner referred to an asymptomatic small solitary nodule on scalp of a 24-year-old male as solitary naevus lipomatosus cutaneous superficialis. Currently the solitary form of NLCS is termed pedunculated lipofibroma.[5,6]

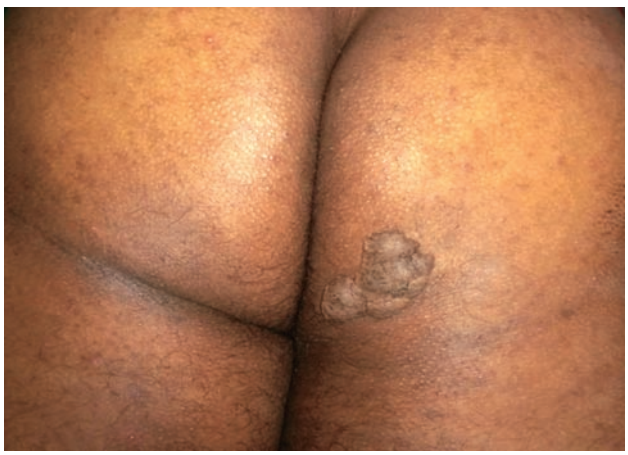

Figure 3. Cerebriform plaque studded with multiple comedo-like lesions over the right buttock.

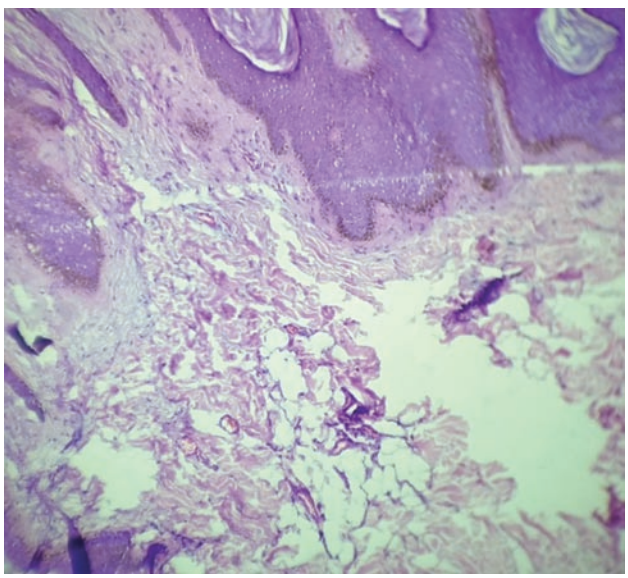

Figure 4. Papillary and reticular dermis showing lobules of adipose tissue in hyalinised stroma (H and E: x10).

The classical form usually presents at birth or develops within the first two decades of life. It manifests with multiple soft, non-tender skin coloured to yellow papules and nodules coalescing into plaques usually in the gluteal, pelvic and lower back regions. It is usually unilateral and may be systematised, linear, zosteriform or along the lines of skin folds in distribution. In our first case, it was zosteriform and in the second case, it was a cerebriform plaque in the gluteal skin fold. Both had been present since birth. The solitary form presents after second decade of life and is seen as solitary dome-shaped or sessile papule on the buttocks and thighs. However it can also be seen in unusual sites like ear, scalp, forehead, back, axilla, arm, nose, and clitoris. There is neither gender predilection nor familial tendency in either form.[2,7,8] There are varied schools of thought regarding its pathogenesis and the exact aetiopathogenesis has not been elucidated. Hoffman and Zuhrelle postulated that deposition of adipose tissue is secondary to degenerative changes in dermal collagen and elastic tissue.[3] While some hypothesise that adipocytes originate from the pericytes of dermal vessels, others attribute it to be a true naevus that resulted from focal heterotopic development of adipose tissue.[9,10] Co-existent pigmentary anomalies in the form of café-au-lait macules, scattered leukoderma, hypertrichosis, capillary haemangioma, comedo-like lesions have been reported.[8] In the second case, comedo-like lesions were present, which is uncommon in the literature.

The differential diagnosis for NLCS includes naevus sebaceous, neurofibroma, naevocellular naevi, acrochordons, lymphangioma, focal dermal hypoplasia. Cutaneous biopsy aids in confirmation of diagnosis. The characteristic histopathological features include the presence of ectopic fat in the dermis. The adipocytes most commonly form aggregates in the perivascular area. They may also be present as solitary adipocytes between collagen bundles.[3] These typical features were present in both cases. Epidermal changes may include mild to moderate acanthosis, basket weave hyperkeratosis, increased basal pigmentation and focal elongation of rete pegs.[11] Additional features in dermis may include increased vascularity and excessive, loose or irregular organisation of the connective tissue.[2] Similar dermal collections of adipocytes may be appreciated in focal dermal hypoplasia and melanocytic naevi. The former is differentiated from NLCS on the basis of absence of collagen in the atrophic dermis and absent skin appendages and the latter contains nests of naevus cells which are absent in NLCS.

NLCS usually has an indolent and asymptomatic course and is not associated with systemic abnormalities or malignant changes. The lesions may extend for many years. The largest size documented to date is 40 cm x 28 cm.[12] Treatment is given mainly for cosmetic concerns. Simple surgical excision is curative and recurrence after surgery is rare.[8] Both our cases were referred for surgical excision.

Conclusion

This report highlighted two cases of the classical form of NLCS which had varying morphological features clinically and characteristic features on histopathology. Although it is a benign condition, its progressive increase in size may affect the patient on cosmetic grounds, hence the importance of early recognition and surgical excision.

References

1. Bancalari E, Martinez-Sanchez D, Tardio JC. Nevus lipomatosus superficialis with a folliculosebaceous component: report of 2 cases. Patholog Res Int 2011; 105973. doi:10.4061/211/105973.
2. Pathirana PW, Ranasinghe A, Perera WMT. Nevus lipomatosus cutaneous superficialis: an unusual presentation. J Diagnost Pathol 2013;8:59-62.
3. Metin A, Calko O, Akpolat N. Nevus Lipomatosus Cutaneous Superficialis (Hoffmann-Zurhelle). East J Med 2001;6:26-8.
4. Rasgdale BD. Tumours with fatty, muscular, osseous, and/or cartilaginous differentiation. Lever's histopathology of the skin, 10th ed. Elder DE, Elentisas R, Jhonson BL, Murphy GF, Xu X editors. Wolters Kluwer-Lippincott Williams & Wikkins; 2009.
5. Hanada K, Takenaka H, Asai J, Ueda E, Katoh N, Kishimoto S. Pedunculated lipofibroma (Hoffmann-Zurhelle) on the palm. Acta Dermatol Venereol 2006; 86:559-60.
6. Dhamija A, Mehedra A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: An unusual presentation. Indian Dermatol Online J 2012;3: 196-8.
7. Gaikwad SL, Sakhare NA, Kumawat UD, Costa GFD. A solitary Nevus Lipomatosus Cutaneous Superficialis: A Rare Case Report. Int J Contemporary Med Res 2016: 3:1760-1.
8. Chougule A, Kumari R, Thappa DM. Giant nevus lipomatosus cutaneous superficialis of the thigh. Indian J Dermatol 2007;52:120-1.
9. Lane JE, Clark E, Marzec T. Nevus lipomatosus cutaneous superficialis. Pediatr Dermatol 2003;20: 313-4.
10. Dotz W, Prioleau PG. Nevus lipomatosus cutaneous superficialis. Arch Dermatol 1984;120:376-9.
11. Buch AC, Paniker NK, Karve PP. Solitary nevus lipomatosus cutaneous superficialis. J Postgrad Med 2005;51:47-8.
12. 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.