Case Report

Naevus lipomatosus cutaneous superficialis: report of two cases

皮膚淺表脂肪性痣的兩例報告

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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.

Keywords: Adipocytes, dermis, hamartoma, naevus

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. The two clinical types recognised are the classical (known as Hoffman Zurhelle) and the solitary (known as pedunculated lipofibroma). 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. The solitary type presents as a solitary dome-shaped papule or nodule.
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.\textsuperscript{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
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 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.\(^5,6\)

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. 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. Additional features in dermis may include increased vascularity and excessive, loose or irregular organisation of the connective tissue. 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. Treatment is given mainly for cosmetic concerns. Simple surgical excision is curative and recurrence after surgery is rare. 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**