
Case report

Nevus lipomatosus cutaneus superficialis: an unusual presentation

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Introduction

Nevus lipomatosus cutaneus superficialis (NLCS) is an uncommon benign hamartomatous condition characterized by ectopic adipose tissue in the dermis. Clinically, it is classified into two types, the classical Hoffman-Zurhelle or the multiple form and the pedunculated solitary form (1). In 1921, Hoffmann and Zurhelle described the first case of NLCS in a 25-year-old man who presented with multiple soft nodules on the gluteal region (2,3).

In 1968, Weitzner reported a 24-year-old Spanish-American male who presented with an asymptomatic small, solitary nodule on the scalp and in which the biopsy was consistent with NLCS. Because the lesion was solitary and located on the scalp rather than the pelvic or gluteal region, Weitzner referred to the lesion as

solitary nevus lipomatosus cutaneus superficialis (2). Currently, the term “pedunculated lipofibroma” is used to denote solitary forms of NLCS (2,3). This case report is of a solitary form of NLCS or pedunculated lipofibroma occurring on the palm.

Case report

A 49-year-old man presented with a gradually enlarging nodule on his right palm which appeared as a blister without any trigger one month ago. Physical examination revealed a pedunculated, solitary, non-tender, soft nodule measuring 13 x12x10 mm. The excision biopsy was submitted for histopathology.

Histopathological examination revealed a slightly acanthotic epidermis with flattened rete ridges. Both the papillary and reticular

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dermis contained scattered lobules of fat cells entrapped between bundles of dermal collagen fibers. The lobules of fat were mainly localized around blood vessels. Increased vascularity was observed in the ectopic dermal fat. The ectopic fat had no connection with the underlying

subcutaneous fat. The pilosebaceous follicles in the dermis were absent (Fig.1 & 2). Based on the clinical and histopathological features, a diagnosis of a solitary or pedunculated form of NLCS was made.

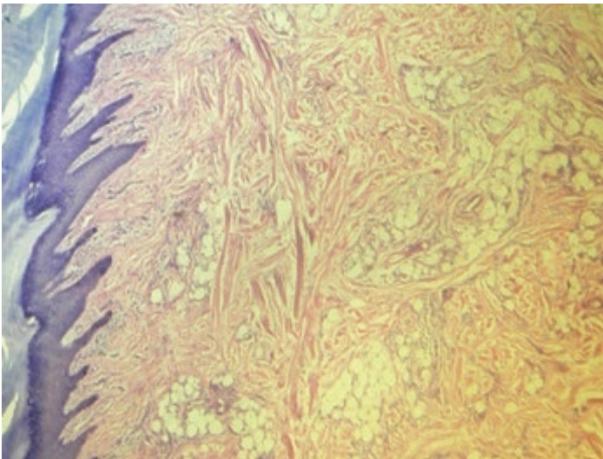


Fig. 1-Ectopic adipose tissue in the papillary and reticular dermis (H&E x100).

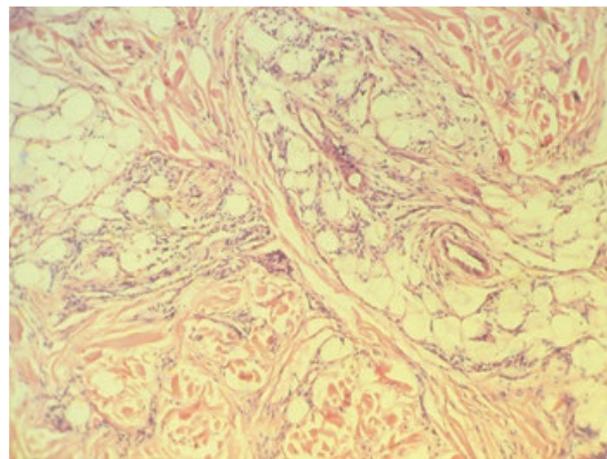


Fig. 2- Scattered lobules of fat cells entrapped between bundles of dermal collagen fibres -arrow (H&E x 400)

Discussion

The two subtypes of NLCS are differentiated based on clinical presentation. The multiple form of NLCS is the Hoffmann-Zurhelle form, or the classic form, characterized by multiple soft, non-tender papules to nodules which commonly coalesce to form plaques. The classic form has a predilection for the gluteal, pelvic and lower back regions. The lesions are present at birth or develop in the first two decades of life (1,2).

The solitary form can present as a single pedunculated or dome-shaped papule to nodule or very rarely a plaque (1,2). Although the solitary form is predominantly found on the buttocks and thighs, it may occur at unusual sites like scalp, axilla, knee, ear, eye and palm as in the present case. Pedunculated lipofibroma is rare in the population and presents after the second decade of life and is

usually seen in the fifth decade of life (2,3). There has been no evidence of correlation with sex or ethnicity (2,4).

The main histological abnormality in either type of NLCS is ectopic fatty tissue in the upper dermis often not connected with the fat of underlying subcutis. The proportion of the dermal fat is variable, ranging from less than 10% of the dermis to over 50% (1,4). When the lesion is small, the fat is usually localized around the subpapillary blood vessels (1). Excessive, loose, or irregular organization of the connective tissue has been noted in many cases. In our case, there was thickening of dermal collagen and the fat was mainly seen around dermal blood vessels as the lesion was small. In NLCS, the blood vessels are frequently increased in number in the upper dermis and ectopic fatty tissue. However in our case, increased vascularity was observed only in the ectopic fatty tissue. In most cases, staining with alcian blue shows substantially increased deposition of mucopolysaccharides in the reticular dermis and fatty tissue (3) which was also evident in our case.

NLCS should be differentiated from nevus sebaceous, focal epidermal hypoplasia, dermal variant of spindle-cell lipoma and other benign papillomas like acrochordons, nevocellular nevi, verrucae, neurofibromas and fibroepi-

thelioma of Pinkus. Histopathological evaluation usually helps in the differentiation. NLCS contains fat cells, but no skin appendages in the dermis.

Nevus sebaceous and other benign papillomas contain skin appendages, but no fat cells in the dermis. Solitary form of NLCS has a broad base when compared to fibroepithelioma of Pinkus. Dermal collections of adipocytes on histopathological examination are also present in old nevocellular nevi and some melanocytic nevi. However, the presence of nevus cells sometimes occupying a small area of the lesion helps in the differentiation. Focal epidermal hypoplasia (Goltz syndrome) also has fat in the dermis, but in this condition there is extreme attenuation of the collagen. The dermal variant of spindle-cell lipoma contains more spindle-shaped cells as well as a fibromucinous stroma.

Treatment for NLCS is not necessary other than for cosmetic reasons (4,8). Systemic abnormalities and malignant changes have not been associated with NLCS. Excision is curative and recurrence after surgery is rare (4,8). In our case, the lesion was completely excised and no recurrences were observed after six months after surgery.

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