A Solitary Nevus Lipomatosus Cutaneous Superficialis: A Rare Case Report

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ABSTRACT

Introduction: Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare benign hamartomatous condition characterized by the presence of ectopic adipose tissue in the dermis. It was first reported by Haffman and Zurhelle in 1921. Clinically, it is classified into two types, the classical- Hoffman-Zurhelle or the multiple form characterized by groups of multiple non-tender, soft, yellowish or skin colored plaques. The other form of NLCS clinically manifest as a solitary pedunculated nodule or sessile papule.

Case report: We report the case of solitary NLCS in 40 years old male presenting as a solitary, pedunculated, soft, non-tender mass measuring 4×3×1cm on the right elbow. Microscopy shows the reticular dermis containing lobules of ectopic fat separating the dermal collagen bundles which suggest the diagnosis of NLCS.

Conclusion: The physicians should be aware of this rare condition because early recognition enables more conservative resection of tumor and less invasive reconstruction of the defect.

Keywords: Nevus Lipomatousus Cutaneous Superficialis, adipocytes, hamartoma

INTRODUCTION

Nevus Lipomatousus Cutaneous Superficialis (NLCS) is an uncommon benign hamartomatous condition characterized by ectopic adipose tissue in the dermis.[1] There is no gender predilection or hereditary predisposition of this disease. Clinically it is classified into two forms. The classical form is characterized by groups of multiple non-tender, soft, pedunculated, yellowish or skin colored papules, nodules or plaques. Classical NLCS is mostly reported to involve the pelvic or gluteal region but can also rarely occur on the abdomen, chest and face.[2]

They are usually present at birth or emerge during the first two decades of life. It was first reported by Haffman and Zurhelle in 1921. The other form of NLCS manifests as a solitary dome shaped or sessile papule on the buttock and thigh.[3] Unusual sites like scalp, axilla, knee, ear, eye, nose and clitoris can also be involved.[1] The solitary form presents after second decade of life. It is also known as pedunculated lipofibroma.[4] In 1968, Weitzner reported a 24 year old Spanish-American male who presented with an asymmetric small, solitary nodule on the scalp and in which the biopsy was consistent with NLCS.[1,2]

We report here a case of solitary form of NLCS occurring on the elbow.

CASE REPORT

A 40 years old man presented with a gradually increasing swelling on the right elbow since 3 months. On physical examination the swelling was solitary, pedunculated, non-tender, soft measuring 4×3×1cm.

The excision biopsy was submitted to the department of pathology for histopathological examination.

Gross: Tissue mass of size 4×3×1cm with external surface covered with thick, wrinkled skin and cut surface whitish yellow. There was no ulceration, pigmentation and hair growth.

Microscopic examination: Hematoxylin and Eosin stained sections of the lesion revealed lining stratified squamous epithelium with flattened rete ridges. Both papillary as well as reticular dermis contain lobules of ectopic fat separating the dermal collagen bundles. The adipose tissue was not encapsulated and was mature. The ectopic fat had no connection with the underlying subcutaneous fat. Fat lobules were mainly localized around the blood vessels with sparse lymphocytic infiltrate. Dermal appendages were absent in the sections studied.

Based on clinical and histopathological features, a diagnosis of solitary NLCS was given.

DISCUSSION

NLCS was first described by Haffman and Zurhelle in 1921. NLCS is a relatively rare disease characterized by groups of ectopic fat cells in the papillary or reticular dermis. Two clinical forms have been identified. The multiple form or the classic form is characterized by multiple soft non-tender skin colored or yellow papules, nodules or plaques usually develop shortly after birth or during the first two decades of life.[3-5]

The classic form has a predilection for the gluteal, pelvic and lower back region.[1] The solitary form can present as a single pedunculated or dome shaped papule or nodule on the buttock and the thigh. Unusual sites like the scalp, axilla, knee, ear, eye, nose, clitoris and palm can also be involved.[1]

The solitary form presents after the second decade of life. There is no gender predilection and patients are otherwise in good health.[5] The main histological abnormality in either type of NLCS is ectopic fatty tissue in the upper dermis often not connected to the fat of the underlying subcutis.[1] The proportion of dermal fat is variable ranging from less than 10% of the dermis to over 50%.[2] In our case ectopic adipose tissue was mainly seen around dermal blood vessels.

Presumably fat cells in the dermis were the result of local heterotopic development of the adipose tissue. NLCS was presumed to be the result of displacement of subcutaneous

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How to cite this article: SL Gaikwad, NA Sakhare, UD Kumawat, GF D'Costa. A solitary nevus lipomatosus cutaneous superficialis: a rare case report. International Journal of Contemporary Medical Research 2016;3(6):1760-1761.

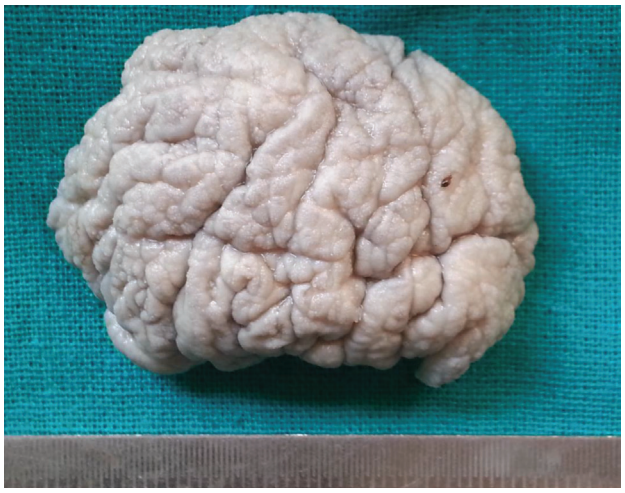

Figure-1: Gross photograph showing solitary nodule with cerebriform surface

Figure-2: Gross photograph showing whitish yellow cut surface

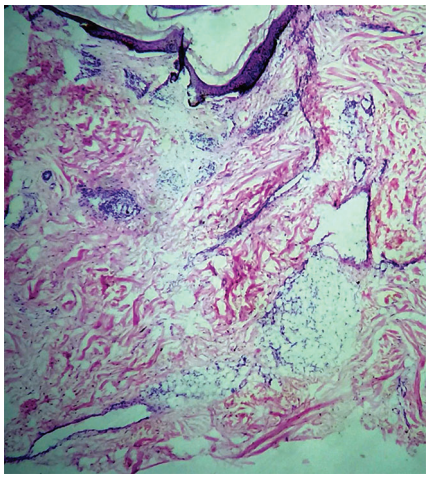

Figure-3: Ectopic adipose tissue in reticular dermis (H and E ×100)

adipose tissue embedded into the dermis. Recently electron microscopic findings strongly confirmed the perivascular origin of young adipocytes and the differentiation into mature fat.[4]

NLCS should be differentiated from nevus sebaceous, fibroepithelioma, nevocellular nevi, focal epidermal hypoplasia and the dermal variant of spindle cell lipoma.

Nevus sebaceous contains skin appendages but no fat cells

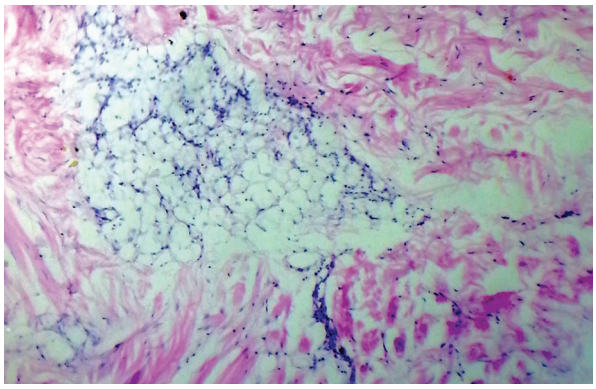

Figure-4: Scattered lobules of ectopic adipose tissue entrapped between bundles of dermal collagen fibers (H and E ×400)

in the dermis. The solitary form of NLCS has a broad base when compared to fibroepithelioma. Dermal collections of the adipocytes are also present in nevi however the presence of nevus cells sometimes occupying a small area of the lesion helps in differentiation.

Focal epidermal hypoplasia also has fat in the dermis along with extreme attenuation of collagen. The dermal variant of spindle cell lipoma contains more spindle shaped cells and fibromucinous stroma.[1]

Another peculiar variant of this condition is marked by excessive, symmetric circumferential folds of skin with underlying NLCS and affect the neck, forearms, lower legs and resolve spontaneously during childhood. It has been described as Michelin tire baby syndrome. This syndrome is inherited as an autosomal dominant trait and is characterized by deletion of chromosome 11.[5]

For cosmetic purpose, surgical excision is the best choice of treatment.[6]

CONCLUSION

The physicians should be aware of this rare condition because early recognition enables more conservative resection of tumor and less invasive reconstruction of the defect.

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Source of Support: Nil; **Conflict of Interest:** None

Submitted: 24-04-2016; **Published online**: 30-05-2016