Solitary And Pedunculated Nevus Lipomatosis Cutaneous Superficialis Of The Thigh– A Rare Case Report And Review Of Literature

ABSTRACT

Background: Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous idiopathic condition. The characteristic feature of this condition is the presence of mature adipocytes in the dermis. Two forms of NLCS are identified clinically vis-à-vis classical form and solitary form. The latter is seen in adults and is a rarer variant than the former.

Case Representation: This case report describes a solitary and pedunculated form of NLCS seen in the thigh of a young adult, a rare variant of NLCS. The swelling was excised surgically followed by good recovery with no recurrence.

Conclusion: Early recognition of this benign condition will result in less morbidity amongst the patients with regards to cosmesis especially when the lesion can recur.

Keywords: Pedunculated nevus lipomatosus cutaneous superficialis, solitary variant, NLCS, lipofibroma

INTRODUCTION

"Hoffman and Zuhrelle in 1921 first described a rare benign hamartomatous idiopathic condition called nevus lipomatosus cutaneous superficialis (NLCS)" [1]. "It is characterized by the presence of ectopic mature adipose tissue within the dermis. Literature describes two clinical variants of NLCS - the classical form, consisting of multiple skin-colored cerebriform, pedunculated nodules that most often combine to form a plaque and the second form which is relatively a rare form, presenting as a solitary sessile papule or nodule usually dome- shaped" [2] Very few cases on solitary form of NLCS have been published in the literature. A solitary, pedunculated neoplasm can be easily misdiagnosed as papilloma or skin tag. This case report describes a solitary and pedunculated NLCS in a young adult, a rare variant of NLCS with review of the literature.

CASE REPORT

A young female aged 27 years presented to our department with a four-year-old history of a swelling on the inner side of left upper thigh. The swelling had originally appeared without any trigger and gradually increased to the present size over the four years. There was no significant family history. Physical examination showed a pedunculated, solitary nodule measuring 40×40 mm. The prominent stalk was 10 mm long. The swelling was skin-colored, nodular in appearance, soft in consistency, non-tender, with no sinuses or ulceration. (Fig 1A) Surrounding skin appeared normal. No other associated swellings were noted. Fine needle aspiration of the swelling reported scattered mature adipocytes with many fibrous strands against a hemorrhagic background. Under local anesthesia, an elliptical incision was marked around the stalk and deepened to completely excise the swelling. (Fig 1B) Incision was closed using non-absorbable sutures (polyethelene 4-0). Histopathological examination (HPE) of the swelling showed infiltration of the epidermis and dermis with chronic inflammatory cell infiltrate. The dermis showed presence of mature adipocytes were seen in lobules separated by fibrocollagenous stroma. Blood vessels were also seen in the stroma. No dysplastic changes present. (Fig 2) The findings were suggestive of NLCS, solitary pedunculated variant. Post-operative period was uneventful. Sutures were removed in two weeks after the surgery. One year follow-up showed no recurrence.

DISCUSSION

Mehregan et al., suggested the term "pedunculated lipofibroma for the solitary form of nevus lipomatosus, in view of its characteristic clinicopathological features".[3] "Pedunculated lipofibroma is a rare, benign connective tissue neoplasm. It is solitary, slow growing and is distinctively diagnosed by the presence of ectopic adipose tissue in the dermis. The histological features are like classic NLCS, which was originally classified into two clinical types, a classic, multiple form, and a solitary form".[1] "In the multiple type of NLCS, the lesions are either congenital or develop during the first three decades of life. The common sites of occurrence are

usually the gluteal region, lower back, and the superior, posterior thighs. The solitary nodular, papular or pedunculated lipofibroma occurs later in life (usually over 20 years of age) and is found in unusual sites like axilla, knee, eyelid, nose, ear/pinna, clitoris, scrotum, or skin of the scalp".[4-6)]

NLCS are slow growing and asymptomatic in nature. However, large lesions tend to ulcerate with superimposing bacterial infections.[2] "Ulnar neuropathy due to compression by a large NLCS has been reported where a partial excision of the lesion was done due to its benign nature".[7] There is no documented genetic predilection, but one study has reported the association of 2p24 deletion.[8]

"The pathogenesis of NLCS is unknown. Various theories have been postulated vis-a-vis degenerative changes in the collagen and elastic tissue resulting in the displacement of subcutaneous adipose tissue into the dermis. Another theory explains origination and differentiation of lipoblasts to adipose cells from the walls of dermal capillary vessels". [9]

"The main histological abnormality in either type of NLCS is ectopic fatty tissue in the upper dermis, distinct from the normal subcutaneous fat. In many cases, the connective tissue has been seen to be organized irregularly. Foci of adipocytes are seen around the dermal blood vessels.[10,11] Staining with alcian blue shows increased deposition of mucopolysaccharides in the reticular dermis and fatty tissue in majority of the cases".[12]

Clinically and histologically, the differential diagnosis for a pedunculated NLCS includes other benign papillomas, including acrochordons, seborrhoeic keratosis, nevocellular nevi, verrucae, neurofibromas, fibroepithelioma of Pinkus, eccrine poroma, focal dermal hyperplasia (Goltz syndrome).[4] HPE will form the key contributor to the final diagnosis.

"The treatment of pedunculated NLCS involves complete excision for both therapeutic and cosmetic reasons. Excision of large sessile plaques may result in the need for skin grafts or a combination of tissue expansion and flap advancement surgery to cover the post-excision defects. Other non-surgical modalities such as cryotherapy, carbon dioxide ablative laser, corticosteroids topical applications and intralesional phosphotidylcholine and sodium deoxycholate injections have been used for the removal of these lesions".[13 - 15] Recurrences have been noted following these modalities when surgical excision becomes an absolute necessity.[14] Although there are no reports in the literature about any malignant transformation of

pedunculated lipofibroma, it is prudent to examine these swellings for malignancy considering the incidence being rare.

CONCLUSION

Nevus lipomatosus cutaneous superficialis is a rare skin pathology and solitary, pedunculated form appears to be a rarer variant. Early recognition of this benign condition will result in less morbidity amongst the patients with regards to cosmesis. Large, pedunculated lesions are susceptible to torsion, ulceration. The comprehensive review of the literature has suggested that surgical excision remains to be the gold standard treatment to prevent recurrence and early intervention will prevent the need for large reconstructive options unless indicated.

STATEMENTS AND DECLARATIONS

Conflicts of interest

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consent

Informed consent was obtained from the patient for the use of photographs for publication. The consent has been formally documented in the medical record.

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that they have no known competing financial interests OR non-financial interests OR personal relationships that could have appeared to influence the work reported in this paper.

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FIGURE LEGENDS

Figure 1A: Pre-operative image of the pedunculated, solitary nodule measuring 40×40 mm with a prominent stalk measuring 10 mm long. The swelling was skin-colored, nodular in appearance, with no sinuses or ulceration.

Figure 1B: Excised swelling with the stalk

Figure 2: Low power view showing hyperkeratosis, irregularly organized connective tissue and presence of mature adipocytes in the dermis.