Case report

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Nevus Lipomatosus Cutaneous Superficialis: A Rare Occurrence

Jyoti Verma[1], Jyotsna N. Bharti[2], Rajesh K. Jarang[3], Rajashekar Mohan[4]

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Abstract

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare cutaneous hamartoma characterized by the ectopic presence of mature adipocytes in papillary and reticular dermis without connection to the subcutis. It usually has a predilection for buttocks, upper thigh and back. It presents itself as skin to yellow colored discrete or confluent papulo-nodular soft, fleshy sessile mass and has two clinical forms described in the literature, classical and solitary types. The classical form is usually composed of multiple and grouped skin-colored, pedunculated nodules. A second and more rare form is characterized by a solitary dome-shaped sessile papule or nodule. Microscopically it presents with the fat deposition in the dermis and is considered to be the consequence of the degenerative changes in the connective tissues. Its pathogenesis is unknown; however, several theories have been proposed. Some attribute it to the adipocyte metaplasia during the degenerative course and some to the development displacement of adipose tissue while others, as a consequence of maturation of adipocytes from mononuclear cells differentiation into lipoblasts in the perivascular zone. Histopathology shows clusters of ectopic presence of adipose fat among the desiccating collagen fibres in the dermis without any connection with the subcutaneous fat. Herein, we present a case of solitary NLCS over left flank associated with left upper thigh acrochordon in a 30 years female.

Keywords: Nevus lipomatosus cutaneous superficialis, hamartoma, ectopic, adipocytes.

Author Affiliation: [1]Assistant Professor, [2]Additional Professor, and Head of Pathology Department, [3]Senior Resident II, [4]Professor and Head of Department of Surgery, Department of Pathology, All India Institute of Medical Sciences, Mangalagiri-522503, Andhra Pradesh, India.

Corresponding Author: Jyoti Verma, Assistant Professor, Department of Pathology, All India Institute of Medical Sciences, Mangalagiri-522503, Andhra Pradesh, India.

E-mail: drjyoti.patho@aiimsmangalagiri.edu.in

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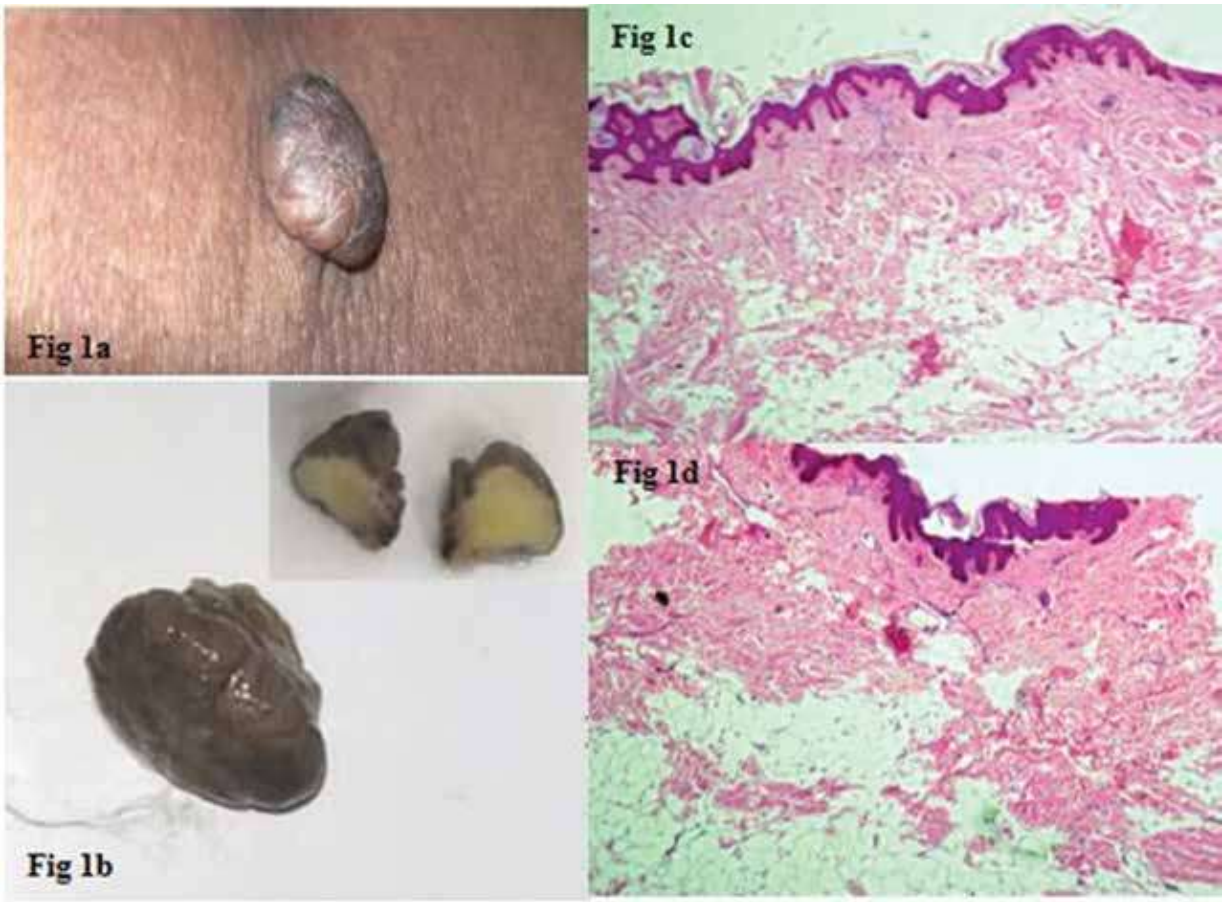
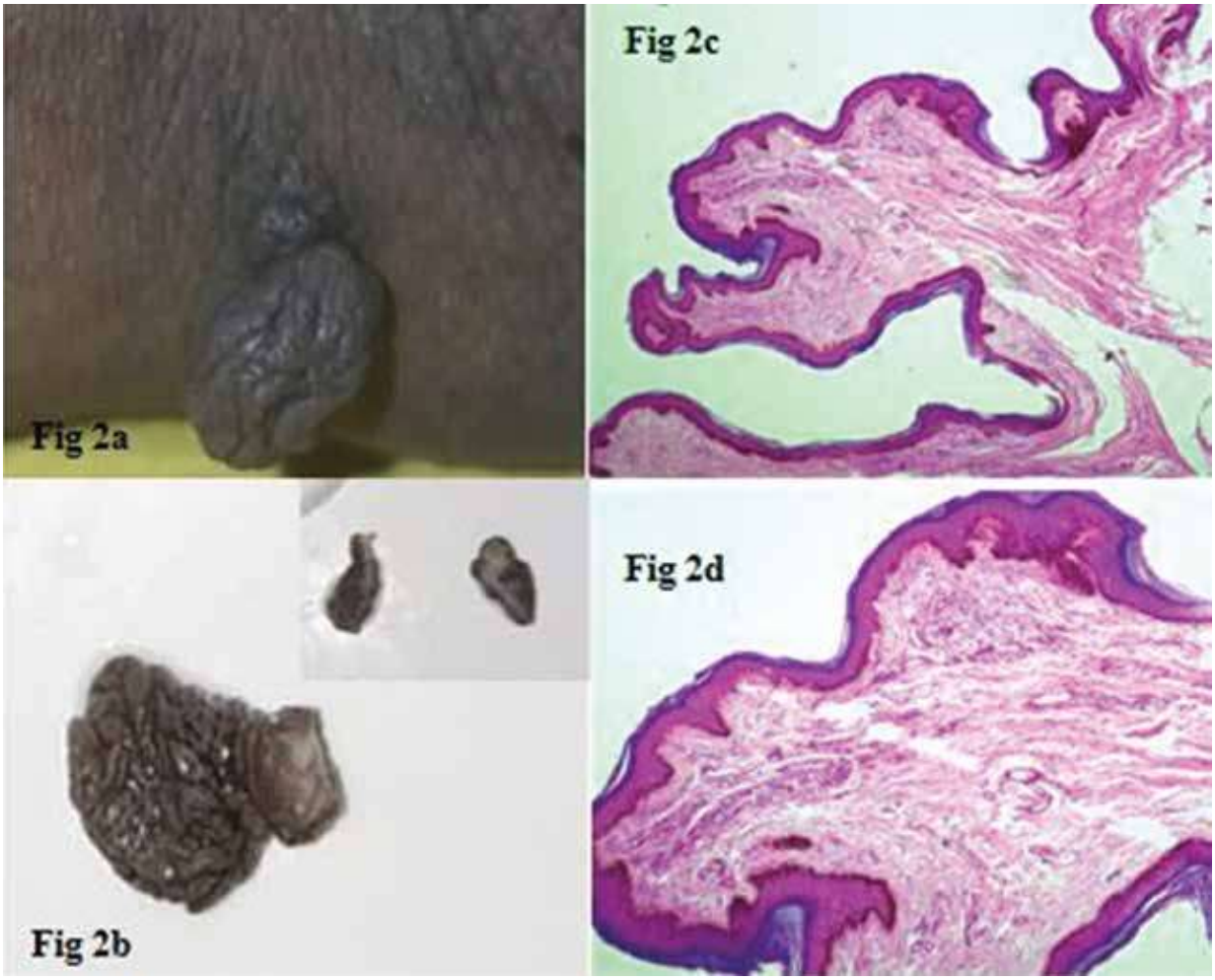
INTRODUCTION

Nevus lipomatosus superficialis is an uncommon, idiopathic, benign, hamartomatous anomaly of the skin. It was first described in 1921 by Hoffmann and Zurhelle as pedunculated lipofibroma.[1,2] It is characterized histologically by aggregates of ectopic mature adipose tissue within the reticular dermis discontinuous with the underlying subcutaneous fat.[3] This malformation is usually present at birth or in early childhood, and there is no sex predilection or hereditary predisposition of this disease. Its preferred sites are buttocks, upper thigh, and lumbar back. Two clinical subtypes described are multiple form (classical type) and solitary form (discrete type). Classical form is the most common with the pelvic-girdle area being the preferential site of involvement. It usually presents at birth or within the first 3 decades of life.[3,4] The second type is the solitary form (rare form) which usually presents as a solitary dome-shaped sessile papule or nodule, typically with late onset and no specific distribution[3]. Histology is characteristic of ectopic, mature adipose tissue in the reticular and papillary dermis.[1,4] We hereby report a case of this rare entity i.e., cutaneous malformation in its solitary form.

CASE REPORT

A 30-year-old female presented to the department of surgery with a complaint of two asymptomatic lesions on the left flank and on the left upper thigh that began to appear for 3 years. On clinical examination, a single, yellowish colored discrete, soft, sessile, round, fleshy, papulo-nodular mass was present near the area of the left flank with cerebriform surface. Another skin-colored lesion with wrinkled appearance was observed in the upper thigh. Both lesions were clinically considered papilloma. There were no associated comedo-like lesions, hypertrichosis, and café-au-lait or hypopigmented macules. Rest of the mucocutaneous examination was not significant. There was no history of a rapid or recent increase in size, pain, or bleeding. There was no family history of similar lesions. Both lesions were surgically excised and the specimens were sent to the department of pathology for histopathological examination (HPE). The specimen received from





the flank area was yellowish-colored polypoid mass with a cerebriform surface measuring 2.0x1x0.8 cm. The cut surface revealed skin with an underlying yellowish white dermis. [Figure 1a-d] The second specimen from the left upper thigh was a skin-covered polypoid mass measuring 1x1x0.5 cm and the cut surface showed gray and white areas. HPE of the first specimen revealed the proliferation of mature ectopic adipocytes in the reticular dermis interspersed between increased dense collagen fibers. The epidermis exhibited hyperkeratosis with elongated rete ridges. Occasional uninvolved adnexal structures were identified. The differential diagnosis includes neurofibroma, sebaceous nevus and connective tissue nevi. The HPE of the second sample showed an epidermis with mild papillomatosis, hyperkeratosis, and irregular acanthosis. The superficial, mid and deeper dermis was fibrotic. The first lesion was validated as Nevus Lipomatosus cutaneous superficialis and the second lesion was validated as fibroepithelial polyp. [Figure 2a-d].

DISCUSSION

Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon, idiopathic, asymptomatic, benign hamartomatous skin lesion, characterized by a collection of mature adipocytes ectopically situated in the dermis and first described by Hoffman and Zurhelle in 1921.[2] Fatty tissue proportion in the dermis varies greatly from < 10% to more than 50%.[5]

NLCS manifests itself usually at birth, but is also known to appear in the later phase of life; there is neither sex predilection nor familial trend in this disorder.[6,7] NLCS may present itself clinically in two forms; The classical lesion or multiple type, first reported by Hoffman and Zurhelle, and the commonest type, consists of multiple, yellowish, skin-colored or flesh colored, soft, sessile, nontender, pedunculated, papules or nodules which may have a tendency to coalesce into plaques with a smooth or cerebriform surface with

a linear, zosteriform, or segmental distribution. These lesions have predilection for lower trunk especially the pelvic girdle and gluteal, sacral, and lumbar regions in a zonal pattern of involvement.[6, 7] It occurs at birth or during first three decades of life.[8,10] The second clinical pattern of NLCS is a solitary form or discrete type, which presents itself as single sessile or pedunculated nodule with a late onset of presentation, usually during third to sixth decades of life, much later than the classical form with no specific distribution or predilection for particular sites, but it has been reported on the arms, knees, axillae, ears, nose, calf, clitoris, and scalp. It clinically appears as a solitary, skin-colored, domed papule or nodule mimicking a skin tag. Index case showed late onset of presentation in (third decade), due to the fact that it was a solitary form.

NLCS overall follows an asymptomatic or static course, however, unusual growth patterns have been reported in literature such as giant NLCS, comedo-like plugs, foul-smelling discharge, and trauma or ischemia associated ulcerated lesions. Also some case reports have described rare coexistent anomalies such as café-au-lait macules and scattered leukoderma with NLCS, angiokeratoma of Fordyce, and hemangioma. Recurrence has rarely been reported. There are no systemic abnormalities or malignant transformation.

Index case of NLCS was coexistent with acrochordon. Fibroepithelial polyp, acrochordons or skin tags are common benign neoplasms of the skin often associated with obesity. These small pedunculated polyps are frequently a nuisance for patients, typically developing around the neck, axilla, and groin areas. Without histological confirmation, acrochordons cannot reliably be distinguished from benign melanocytic nevi and neurofibromas. However, since all of the possible differentials are benign, pathological confirmation is not typically necessary since it will not impact management protocol.

There are certain theories put forward to explain the heterotopic presence of adipose tissues within the dermis without any continuity to the subcutis. It may originate from the precursor cells from the dermal vessels or pericytes by mesenchymal perivascular differentiation of lipoblasts, and adipose metaplasia in dermal connective tissue as in fetal lipogenesis. Few have proposed deletion of chromosome 2 (2p24) as its etiology.[9] However the exact etiopathogenesis is still unknown.

On HPE, there is presence of ectopic aggregates of lobules or dispersed well-differentiated adipocytes intermingled with collagen bundles. Clinical differentials include plexiform neurofibroma, skin tag, fibroepithelial polyp, lymphangioma, and leiomyoma cutis. These can be distinguished histopathologically[10] There is perivascular infiltration of the dermis and subcutis with chronic inflammatory cell infiltrate. Few cases with rare histopathological features have also been reported in literature showing focal elongation of rete pegs with increased basal pigmentation, reduced number of adnexal structures and abnormal presence of folliculo sebaceous units for ex: Sebaceous trichofolliculoma, folliculo sebaceous cystic hamartoma, and dermoid cysts[11] Our case showed mature adipocytes in the dermis with collagen bundles interspersed between these adipocytes.

Treatment of choice is surgical excision and is usually for cosmetic reasons if indicated, otherwise it is curative[12] Cryotherapy and carbon dioxide laser has also shown partial improvement[13] Malignant transformation and recurrences after surgery have not been reported so far.

CONCLUSION

This report emphasizes the importance of early diagnosis of this rare neoplasia, which although benign, can grow to a large size and may cause aesthetic concern to the patient. Lesions are generally non-progressive and systemic abnormalities and malignant alterations have not been associated with this abnormality.

ABBREVIATIONS

HPE: Histopathological examination.

NLCS: Nevus lipomatosus cutaneous superficialis.

DECLARATIONS:

Ethics approval and consent to participate: This case report was conducted by the fundamental principles of the Declaration of Helsinki.

Consent for publication: Verbal consent for the publication and any additional related information was taken from the patient involved in the study.

Availability of data and materials: All the data regarding the findings are available within the manuscript.

Competing interests: The authors declare that they have no competing interests.

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