

Nevus Lipomatosus Cutaneous Superficialis with Spindle Cell Lipoma-like Areas: A Case Report

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ABSTRACT

Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare occurring cutaneous hamartoma with presence of mature adipocytes in dermis. Though first reported in 1921 by Haffman and Zurhelle, fewer cases have been documented in literature since then. Authors, hereby, report a case of nevus lipomatosus cutaneous superficialis in an unusual age of 55-year-old male with multiple painless ill defined sessile nodules of 1.5 to 3 cm on lower back. Surgical excision was performed and histopathology revealed characteristic features as that of NLCS along with intervening areas of spindle cell lipoma. Co-existence of other lesion along with NLCS, both clinically or on histopathology is further an infrequent occurrence. No such case has been reported in literature so far.

Keywords: Benign adipocytic tumours, Hamartoma, Lower back region, Nodule

CASE REPORT

A 55-year-old healthy male presented to Surgery Outpatient Department (OPD) with painless, slow growing multiple nodules on right side of lower back region, present for the past three years. The nodules were non tender, non pruritic and were not associated with any bleeding, ulceration, discharge. No similar lesion was present at any other site. Furthermore, there was no family history of any such lesion.

On clinical examination, there were multiple skin coloured, soft, ill-defined, sessile subcutaneous nodules over the lower back region, ranging in size from 1.5 cm to 3 cm in diameter. The nodules were not fixed with underlying structures. The lesion was resected with ellipse of skin around it and sent for histopathological examination.

On gross examination, the whole specimen measures 11×5×3 cm. Cut surface of the nodules was solid, smooth with homogenous yellowish white areas [Table/Fig-1a,b]. No haemorrhage, cyst or necrosis were identified. Multiple sections were taken from the nodules including the skin.



[Table/Fig-1]: (a) Gross specimen with multinodular subcutaneous swelling; (b) Cut surface showed ill-defined yellowish white solid lesion.

On microscopy, Haematoxylin and Eosin (H&E) stained section showed unremarkable epidermis with an ill defined lesion in reticular dermis, composed of variable sized lobules of mature adipocytes separated by dense fibrocollagenous bands [Table/Fig-2]. The lobules show admixture of mature adipocytes alternating with myxoid areas enriched with spindle cells of bland morphology [Table/Fig-3,4]. Many linear blood vessels and adnexal appendages were also seen in between the lobules. No lipoblast, atypical cell/ necrosis, mitosis were found. After considering various benign adipocytic lesions and variants of lipoma, a diagnosis of nevus lipomatosus cutaneous superficialis with spindle cell lipoma-like areas was rendered. No recurrence was seen during the three months follow-up period.

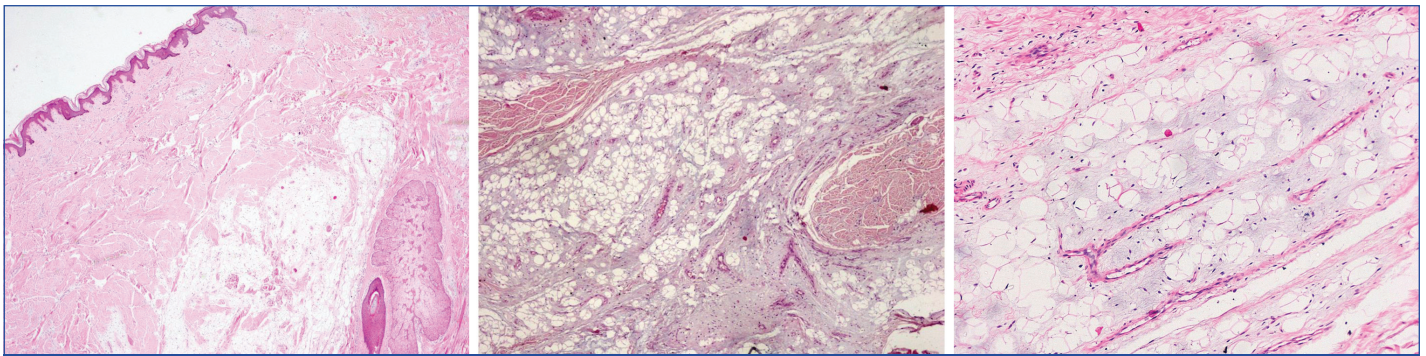
DISCUSSION

Benign adipocytic tumors are fairly common, although rarity is not an exception among few of the variants. Nevus Lipomatosus Cutaneous Superficialis (NLCS) is one such rarely occurring cutaneous hamartoma, characterised by the presence of mature adipocytes in papillary and reticular dermis [1,2]. Co-existence of other adipocytic/non adipocytic lesion along with NLCS is even a rarer occurrence. So far, cases of giant NLCS with multiple folliculosebaceous cystic hamartomas and dermoid cysts and NLCS with localised scleroderma, like appearance has been reported in medical literature [3,4]. The authors report an unusual case of NLCS which, in addition, had areas of spindle cell lipoma.

Hoffmann E and Zurhelle E, were the first to describe NLCS in 1921 [5]. This uncommon condition occur as classical (multiple) and solitary types, clinically. Both the types have similar clinical presentation and differ only in terms of age, number and site of lesion. Classical forms usually present as multiple, yellowish to skin-coloured, sessile/pedunculated, papules/nodules/plaques in a linear, zosteriform, or segmental pattern at birth or in the first two decades of life over pelvic girdle, gluteal region, lower trunk and upper thigh [6]. On the contrary, solitary form often appears in third and sixth decades of life as an asymptomatic, yellowish to skin-coloured, solitary, papule/nodule with no predilection for any particular site [7]. These lesions usually remain static but can gradually progress in size over years. Uncommonly, these lesions may ulcerate [8]. Clinically, NLCS can be mistaken as neurofibromatosis, lymphangioma, haemangioma, fibroepithelial polyp [9]. Both the forms show similar histomorphological features and surgical excision is the main modality of treatment [6, 10].

Characteristic histopathological feature of NLCS is abnormal presence of mature adipocytes or their aggregates in the dermis, especially around blood vessels or eccrine glands [6]. This ectopic fat may variably occupy 10-70% of the dermis [11,12]. These adipocytes may or may not be separated from subcutaneous fat. A higher proportion of adipose tissue can be noted in larger lesions as compared to smaller lesions [12].

The epidermis remains normal but can be attenuated to acanthotic [12]. Increased vascularity and perivascular mononuclear cell infiltration in dermis has been reported in NLCS [13]. Also, uncommonly, adnexal structures can be reduced in number and may show perifollicular fibrosis [14]. In present case, the adipocytes comprised of 60% of the dermis, while epidermis and dermal appendages were unremarkable.



[Table/Fig-2]: Microscopy showed lobules of mature adipocytes extending into reticular dermis (H&E, 2X). **[Table/Fig-3]:** Lobules of mature adipocytes with admixed spindle cells and myxoid areas (H&E, 4X). **[Table/Fig-4]:** Mature adipocytes with admixed bland spindle cells and prominent myxoid areas (H&E, 10X). (Images from left to right)

On the other hand, spindle cell lipoma is also an infrequent histological variant of lipoma, usually presents as subcutaneous lesions of mature adipocytes. It usually arises on posterior neck, upper back and shoulder of elderly males between the 5th and 7th decade of life and are composed of mature adipocytes, bland spindle cells and collagen bundles in varying proportions [15,16]. Uncommonly, NLCS can also show spindle cells representing immature adipocytes focally [12]. In present case, spindle cells comprised almost 35% of the lesion.

Case reports of NLCS with co-existing conditions like café-au-lait macules, leukoderma, hypertrichosis, folliculo-sebaceous cystic hamartomas, dermoid cysts has been reported in literature [3,4,17]. Shiomi et al., reported a case of nevus lipomatous cutaneous superficialis with spindle cell lipoma-like histopathological features. [18]. Another unpublished observation by Rajaji et al., mentions a case of NLCS with features of spindle cell lipoma [19]. But its existence with any other adipocytic lesion or any of its variants has not been seen.

Various theories have been postulated for the origin of dermal adipocytes in NLCS. These have been suggested to arise from pericytes, or from perivascular mesenchymal tissue [20,21]. Hoffmann E and Zurhelle E, suggested that the deposition of adipose tissue in dermis is a result of degenerative change in the connective tissue [4]. Another study reported the deletion of 2p24 in NLCS, indicating a role of genetic factors in development of NLCS [22]. Still, the aetiopathogenesis of NLCs remains unclear.

CONCLUSION(S)

The present case report highlights one of the rare and interesting presentation of nevus lipomatous cutaneous superficialis in terms of age, site and its co-existing histopathological features of spindle cell lipoma. Both come under the category of rare occurring adipocytic lesion. Surgical excision is the mainstay of treatment. Rare recurrences and complications have been reported, though early diagnosis enables more conservative resection.

REFERENCES

- Patil SB, Narchal S, Paricharak M, More S. Nevus lipomatous cutaneous superficialis: A rare case report. *Iran J Med Sci.* 2014;39:304-07. PMID: 24850990; PMCID: PMC4027012.
- Bancalari E, Martínez-Sánchez D, Tardío JC. Nevus lipomatosus superficialis with a folliculosebaceous component: Report of 2 cases. *Patholog Res Int.* 2011;18:105973.
- Brasanac D, Boricic I. Giant nevus lipomatosus superficialis with multiple folliculosebaceous cystic hamartomas and dermoid cysts. *J Eur Acad Dermatol Venereol.* 2005;19:84-86.
- Ioannidou DJ, Stefanidou MP, Panayiotides JG, Tosca AD. Nevus lipomatosus cutaneous superficialis (Hoffmann-Zurhelle) with localized scleroderma like appearance. *Int J Dermatol.* 2001;40:54-57.
- Hoffmann E, Zurhelle E. Ubereinen nevus lipomatodes cutaneous superficialis der linkenglutaaliegend. *Arch Dermatol Syph.* 1921;130:327-33.
- Dhamija A, Meherda A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: An unusual presentation. *Indian Dermatol Online J.* 2012;3:196-98.
- Bhushan P, Thatte SS, Singh A. Nevus lipomatosus cutaneous superficialis: A report of two cases. *Int J Dermatol.* 2016;61:123.
- Dhar S, Kumar B, Kaur I. Naevus lipomatosus superficialis of Hoffman and Zurhelle. *Indian J Dermatol Venereol Leprol.* 1994;60:39-40.
- Pujani M, Choudhury M, Garg T, Madan NK. Nevus lipomatosus superficialis: A rare cutaneous hamartoma. *Indian Dermatol Online J.* 2014;5:109-10.
- Ranjikesh MR, Herizchi QH, Yousefi N. Nevus lipomatosus cutaneous superficialis: A case report with histologic findings. *J Turk Acad Dermatol.* 2009;3:01-03.
- Umashankar T, Prasad T, Rajeshwari SH. Naevus lipomatosus superficialis: Clinicopathologic study of a case. *Indian J Pathol Microbiol.* 2003;46:444-45.
- Dhamija A, Meherda A, D'Souza P, Meena RS. Nevus lipomatosus cutaneous superficialis: An unusual presentation. *Indian Dermatol Online J.* 2012;3:196-198.
- Buch AC, Panicker NK, Karve PP. Solitary nevus lipomatosus cutaneous superficialis. *J Postgrad Med.* 2005;51:47-48.
- Takashima H, Toyoda M, Ikeda Y, Kagaura M, Morohashi M. Nevus lipomatosus cutaneous superficialis with perifollicular fibrosis. *Eur J Dermatol.* 2003;6:584-86.
- Fletcher CD, Martin-Bates E. Spindle cell lipoma: a clinicopathological study with some original observations. *Histopathology.* 1987;11:803-17.
- Enzinger FM, Harvey DA. Spindle cell lipoma. *Cancer.* 1975;36:1852-59.
- Khandpur S, Nagpal SA, Chandra S, Sharma VK, Kaushal S, Safaya R. Giant nevus lipomatosus cutaneous superficialis. *Indian J Dermatol Venereol Leprol.* 2009;75:407-08. 10.4103/0378-6323.53149.
- Shiomi T, Matsuno M, Nishimura H, Moriya T. Coexistence of three nevus lipomatosus cutaneous superficialis with typical, cutaneous adenolipoma-like, and dermal spindle cell lipoma-like histopathological features in a patient. *J Cutan Pathol.* 2021;48:961-64.
- https://cdn.ymaws.com/www.aocd.org/resource/resmgr/meeting_resources/2018Spring/Syllabus/Posters/Rajaji.pdf. (Last accessed on 10-3-2022).
- Pai VV, Naveen KN, Athanikar SB, Rao R, Sori T. Unusual presentation of naevus lipomatosus cutaneous superficialis. *Int J Dermatol.* 2013;58:486-87.
- Stewart CL, Novoa RA, Seykora JT. Tumors of the epidermal appendages. In: Elder DE, Walter Lever, editors. *Lever's histopathology of the skin.* 11th ed. Philadelphia: Wolters Kluwer. 2015:1062-63.
- Cardot-Leccia N, Italiano A, Monteil MC, Basc E, Perrin C. Naevus lipomatosus superficialis: A case report with a 2p24 deletion. *Br J Dermatol.* 2007;156:380-81.

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