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Short Communication

Nevus Lipomatosis Cutaneous Superficialis – A clinicopathologic study of the solitary type

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ABSTRACT

Background: Nevus Lipomatosis Cutaneous Superficialis (NLCS) is a rare, benign hamartomatous lesion characterized by the ectopic presence of mature adipocytes in the reticular dermis not associated with the underlying subcutaneous tissue. Two clinical forms-classical and solitary occur. The solitary form is relatively uncommon, due to which these lesions are commonly clinically misdiagnosed. The aim was to study the clinical and histopathological features of the solitary type of NLCS.

Methods: Seven cases of histopathologically documented solitary type of NLCS which presented at our institute between August 2013 and June 2014 were retrospectively analysed for clinical data and histopathological findings. Haematoxylin and Eosin (H&E) and Elastic Van Gieson (EVG) stained slides were studied in all cases.

Results: Mean age of the patients was 42.5 years. These lesions were more common in adult females (5/7). Thigh was the commonest location (4/7). The mean duration of these lesions was 2.0 years. Clinical diagnosis was papilloma (4/7) and acrochordon (3/7). Histopathology revealed the presence of varying amounts of mature ectopic adipocytes in the dermis located around dilated, ectatic blood vessels. Disorganised dermal collagen bundles and atrophic pilosebaceous units were seen.

Conclusion: This study is a first from the Indian subcontinent and highlights the need for awareness of this rare clinical condition both by the dermatologists and the surgeons. Histopathology is essential for diagnosis as clinically they may mimic papillomas or skin tags. An early diagnosis may permit a more conservative resection of the tumour.

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Introduction

Nevus Lipomatosis Cutaneous Superficialis (NLCS) is a rare benign hamartomatous condition characterized by the presence of mature ectopic adipocytes in the dermis, first described by Hoffmann and Zurhelle in 1921. Clinically it is classified into two forms. The classical form, is commoner characterized by multiple, soft, nontender, cereberiform, pedunculated, yellowish or skin coloured papules, nodules or plaques. The solitary form is uncommon and presents as a single sessile or dome shaped papule.¹ Though there have been many isolated case reports published of NLCS, a study of case series have been very few and most of them describe the classical variant as the predominant type.^{2,3} We highlight the clinicopathological features of 7 cases of the solitary type of NLCS, all of which were clinically misdiagnosed. This calls for a greater awareness of health care providers in clinically recognizing this rare benign condition.

Materials and methods

This retrospective study included all patients' with histopathologically documented NLCS who presented to a multi-speciality hospital over a span of ten months (August 2013 to June 2014). Approval for this study was taken from the Institute Academic committee for analysing the clinical records and histopathological findings of all the patients' included in the study. The clinical data was recorded from the accompanying case sheets. Haematoxylin and Eosin (H&E) slides were reviewed in all cases. In addition, Elastic Van Gieson (EVG) staining was done to study any connective tissue abnormalities.

Results

The clinical profile of all the patients' is given in Table 1. Patients commonly presented to the surgical department (5/7). 71.4% (5/7) were adult females and the thigh was the commonest location (4/7). Mean age of the patients was 42.5 years and the mean duration of the lesions was 2.0 years (range 6 months–4 years). The size was seen to vary from 0.5 cm to 5 × 3 cm. The clinical diagnosis in 4/7 cases was papilloma whereas in 3 cases it was an acrochordon (skin tag). There was no co-existence of the classical type of lesions or

any other anomaly in any of the patients. None of the patients' reported a familial tendency for the same.

Gross examination revealed skin covered sessile lesion without any attached pedicle. Cut section was soft, having a homogenous yellow appearance. Histopathological examination revealed an atrophic epidermis in all cases with one patient in addition exhibiting a mild spongiosis. The dermis in all cases revealed the presence of ectopic adipocytes in varying proportions from 40 to 70%. The blood vessels in the dermis appeared to be increased in all cases. At places, a large central ectatic vessel was surrounded by smaller calibre microvessels. Collagen bundles were seen to be disorganized in 6/7 cases. However, elastic fibres were normal and pilosebaceous units atrophic in all patients' studied. In addition, a mild perivascular chronic inflammatory mononuclear infiltrate was seen in 3 cases.

Discussion

NLCS was first described by Hoffmann and Zurhelle in 1921.¹ The classical form can be present at birth but usually appears in the first two decades of life. It is seen to have a linear, zonal distribution along natural cleavage lines of the skin and has a predilection for the pelvic girdle, sacral and lumbar regions. The solitary form usually appears around the fifth decade and has a predilection for the trunk, though any site may be involved.⁴ Though no familial or sex predilection has been noted, 71.4% (5/7) of our cases were seen in adult females, the mean age of patients' at the time of presentation was younger (42.5 years) and the commonest location was the thigh (4/7).

Most published case series describe patients' with the classical form of NLCS^{2,3} whereas only a single study till date describes a preponderance of the solitary type.⁵ Even though both forms appear insidiously, the classical form is easy to diagnose, however the solitary form is commonly misdiagnosed clinically as seen in our study and corroborated by other workers.³ Our study indicated that smaller lesions were commonly misdiagnosed as acrochordon (skin tag) and the larger ones as papilloma. Some authors have referred to the solitary form as 'pedunculated lipofibromas'.⁶

The lesions grow slowly in size but become stable with time. The size of the lesions in the present series varied from 0.5 cm to the largest measuring 5 × 3 cm. Rarely, huge giant forms of NLCS have also been reported measuring upto 40 × 28 cm.⁷ The cut surface in all cases appeared homo-

Table 1 – Clinical profile of patients' of solitary type of NLCS.

S No	Age/Sex	Location	Duration	Size	Referring department	Clinical diagnosis
1.	56/M	Skin tag in anal cleft	6 month	0.5 cm	Surgery	Acrochordon
2.	46/M	Anterior aspect of right thigh	3.5 years	5 × 3 cm	Surgery	Papilloma
3.	32/F	Papillary growth left thigh	1 year	1.5 × 1 cm	Surgery	Papilloma
4.	35/F	Medial aspect of left thigh	3 years	4 × 3 cm	Dermatology	Papilloma
5.	41/F	Right buttock	1 year	1.5 × 1.5 cm	Surgery	Acrochordon
6.	43/F	Left upper thigh	2 years	4 × 2 cm	Surgery	Papilloma
7.	39/F	Left buttock	4 years	3 × 2 cm	Dermatology	Acrochordon

Fig. 1 – A – Gross image of case 2 of NCLS showing yellowish nodular tumour measuring 5 × 3 cm. B – High power view (×400) revealing central ectatic vessel surrounded by smaller calibre vessels giving a glomeruloid appearance. C – Low power view (×100) of NCLS showing presence of ectopic mature adipocytes in superficial dermis.

genously yellow and was of a softer consistency in contrast to a papilloma which is more firm and whitish (Fig. 1).

NLCS has been reported to be associated with co-existent anomalies like café-au-lait macules and leukoderma, angiokeratoma of Fordyce, trichofolliculoma, and lipedematous scalp.^{2,8} None of our patients' however, reported any co-existent anomalies. These lesions, occasionally may have a foul smelling discharge and may show the presence of comedo-like plugs.¹

Histopathological examination of NLCS revealed an atrophic epidermis in all cases and focal spongiosis in one case. Changes like mild to moderate acanthosis, basket weave hyperkeratosis, increased basal cell pigmentation, focal elongation of rete ridges have been described but were not seen in our cases.⁹

The proportion of ectopic adipocytes in the dermis varied from 40% to 70%, with the larger lesions demonstrating a greater amount of fat than smaller lesions. When only a small amount of fat was present, it was characteristically seen localised around subpapillary blood vessels. The blood vessels appeared ectatic, with a central large vessel surrounded by many smaller calibre daughter vessels giving a 'glomeruloid appearance'. Though ectatic blood vessels have been noted, this glomeruloid appearance has not been described till now (Fig. 2).

Connective tissue abnormalities like thickening of collagen bundles, as reported by other authors were not observed, rather the collagen in the dermis appeared to be loose, disorganised with the fibres running vertically and haphazardly in 6/7 cases in contrast with their parallel orientation seen normally. This irregularity in collagen bundles has been noted by some authors.⁹ A possible role of the collagen bundles in trying to limit the progression of the lesion seems plausible and needs to be considered. Elastic tissue fibres in the dermis did not show any evidence of reduction or degeneration. Pilosebaceous units appeared to be atrophic in all cases (7/7) in

Fig. 2 – Low power view (×100) of NCLS case 5 with hyperkeratosis and disorganized collagen bundles in dermis.

congruence with other studies. However, some authors have also described associated sebaceous trichofolliculoma, folliculosebaceous cystic hamartoma.²

NLCS needs to be distinguished from focal dermal hypoplasia which shows extreme attenuation of collagen in the dermis along with the presence of fat cells. Goltz syndrome is an X-linked disorder, associated with the presence of congenital anomalies. Fibrolipoma shows a considerable proportion of fibrous connective tissue besides dermal adipocytes. Melanocytic nevi, in addition to the presence of nevus cells may show the presence of fat cells which increase with age, representing an involutionary phenomenon.

The pathogenesis of NLCS remains unknown and various theories have been proposed. Though Hoffmann and Zurhelle postulated that the fat deposition in the dermis was secondary to degenerative changes in the connective tissue,¹ we did not observe any degenerative changes in collagen or elastic fibres. Some authors have suggested that the fat cells represented a true nevus which resulted from focal heterotopic development of adipose tissue during embryonic life.⁹ The perivascular location of adipocytes in close association with capillaries as shown by electron microscopic studies suggest an origin from pericytes and has been postulated by other workers.¹⁰ Our findings also seem to find favour with this view as the adipocytes were in intimate association with dilated, ectatic capillaries which appeared to be increased in number. A recent report documents a 2p24 deletion in NLCS and needs to be corroborated further.²

Treatment is usually for cosmetic reasons and surgical resection appears to be adequate. For larger lesions, wide excision with skin grafting remains the treatment of choice. Systemic abnormalities or malignant change have not been reported with NLCS and post surgical recurrence is rare.¹

Conclusion

This is the first study from India reporting a series of 7 cases, all of the solitary type of NLCS in a short span of ten

months. Solitary type of NLCS was more common in adult females and the thigh appeared to be the commonest site. None of these patients' had a correct preoperative clinical diagnosis. The larger lesions were clinically diagnosed as papilloma whereas the smaller ones as an acrochordon. Histopathology revealed the presence of varying amounts of mature ectopic adipocytes in the dermis which were characteristically located around dilated, ectatic blood vessels. Other abnormalities included disorganised dermal collagen bundles and atrophic pilosebaceous units.

This study highlights the need for awareness of this rare clinical condition both by dermatologists and surgeons and histopathological examination is mandatory for a final diagnosis. An early diagnosis may permit a more conservative resection of the tumour. Going ahead, further studies are needed to ascertain the role of genetic abnormalities or even other environmental factors like presence of insulin resistance or metabolic syndrome in the pathogenesis of these lesions in the Indian population. Since this study was a retrospective one, the authors were limited by the lack of anthropometric data, and results of biochemical investigations to enable a conclusive association of the same.

Conflicts of interest

All authors have none to declare.

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