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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, ectactic 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 multispeciality 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 ectactic 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