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Nevus Lipomatosus Cutaneous Superficialis: A Case Series in a Tertiary Care Centre in Garhwal Region

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ABSTRACT
Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous skin lesion characterised by the ectopic presence of mature adipose tissue among the collagen bundles of the dermis, without any connection with underlying subcutaneous fat. It has been a rare entity which presents to a pathologist with invariably inconsistent clinical diagnosis and in our study none came to the pathology department with a provisional clinical diagnosis of NLCS. This case series reports 7 cases and studies the clinical and various histopathological features of the patients of NLCS.

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartomatous skin lesion characterised by the ectopic presence of mature adipose tissue among the collagen bundles of the dermis, without connection with underlying subcutaneous fat. It was first reported by Hoffman and Zurhelle in 1921[1]. It presents clinically as two distinct forms: the more common classical form characterised by grouped soft fleshy skin coloured or yellowish nodules with invariably wrinkled surface and a less common solitary form which presents as a single rounded protrubance or sessile nodule and may have smooth or wrinkled and cerebriform surface[2]. There is a long list of differential diagnosis of NLCS which range from benign non serious lesions to some soft tissue tumours and hence it's pretty much important to rule out other clinically and histologically similar lesions. In this study we report the clinical features along with a detailed account of the histological features of the 7 cases of NLCS.

Case series: All the seven cases with diagnosis of NLCS, spread over a period of 9 years from January 2014 to December 2022, were managed in Veer Chandra Singh Garhwali Government Institute of Medical Science and Research, Srinagar, Garhwal, Uttarakhand and were studied in detail. None of the cases came to the pathology department with a clinical diagnosis of NLCS and other provisional diagnosis were ascribed to these cases.

Case 1: A 53 year old male presented with a mass of 6x2 cm in the right thigh region. The patient was complaining of pain and discomfort in sitting posture. The overlying skin was cerebriform and was clinically suspected to be a case of papilloma. Surgical excision of the lesion was done and the sample was sent for histopathological examination.

Case 2: A 3 year old female presented with multiple sessile nodules close to each other on the gluteal region and altogether measuring 3x1cm in size. The patient was asymptomatic and was clinically suspected to be a fibroepithelial polyp. The surgical excision was done and the sample was sent for histopathological examination.

Case 3: A 27 year old female had a mass of 2.5x1.5 cm on the thigh region which have grown insidiously over period of years. It was asymptomatic and was clinically suspected to be a lipoma. The surgical excision was done and the sample was sent for histopathological examination.

Case 4: A 65 year old female presented with an asymptomatic mass measuring 2.x1.5 cm in the axillary region. FNAC of the lesion was performed and provisional diagnosis of lipoma was ascribed. Surgical excision of the lesion was done and the sample was sent for histopathological examination.

Case 5: A 2 year old boy presented with small multiple closely arranged lesions together measuring 1x2.0cm mass on lower back, since birth. The lesion was suspected to be soft tissue tumour? and surgical excision was performed and sent to histopathological examination.

Case 6: A 17 year old boy with a 1x0.5 cm mass in external auditory canal. The lesion was suspected to be of papilloma and surgical excision was performed.

Case 7: A 38 year old female presented with a history of asymptomatic mass on the right arm measuring 2x3 cms and progressively increasing in size from last 6 months. The lesion had a cerebriform overlying skin and was clinically suspected to be a neurofibroma. Surgical excision was done and sent for histopathological examination.

Histopathological examination of all the 7 cases revealed the features of Nevus lipomatosis cutaneous superficialis and none came with a provisional diagnosis of the same.

A total of 7 cases were studied and out of which 3 (42.85%) were males and 4 (57.15%) were females. Classical form of NLCS, presenting as multiple skin coloured nodules were seen in 2 (28.57%) cases while in 5 (71.43%) cases solitary nodular form was seen. Youngest patient in the case series was a 2 year old boy and the oldest was a 65 years old female. 2 patients had this lesion since birth while the rest developed the lesion in later part of life. Thigh was the most common site of the lesion followed by gluteal region. The size of the hamartoma ranged from 1cm-6 cm in their greatest dimension. Among all those lesions, 3 (42.85%) were less than or equal to 2 cms and 4 (57.15%) were more than 2 cms size in the greatest dimension. In 2 (28.57%) cases the lesions were covered with smooth surfaced skin while in 5 (71.43%) cases they had overlying cerebriform skin. Histopathological changes of all the cases revealed a common finding of presence of mature adipose tissue in the dermis without continuation with subcutaneous fat (fig.1). Flexible amounts of mature adipose tissue were seen in the dermis, varying from nearly 20% to more than 70%, in different cases. The consistent

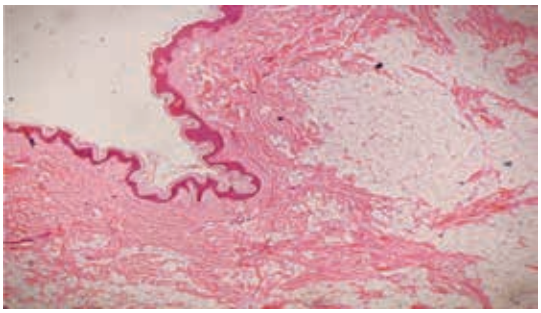

Fig. 1: Microscopic examination showing mature adicpoctes in dermis(H and E stain 40X)

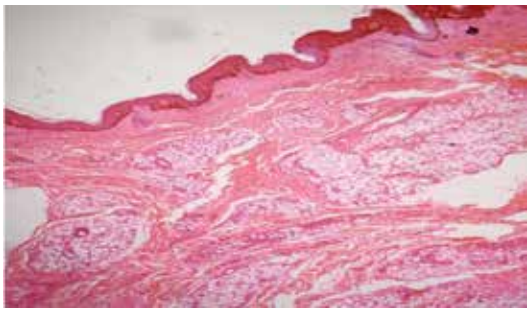

Fig. 2: Microscopic examination showing mature adicpoctes in dermis infiltrated by mononuclear cells(H and E stain 40X)

anomaly seen in the connective tissue components of the dermis was thickened collagen bundles and were seen in 5 (71.42%) cases and mainly restricted to lower dermis. Vascularity was seen increased in the dermis in 3 (42.85%) cases and the increased vascularity was in the form of small arteries, veins and ectatic vessels. Pilosebaceous follicles were seen decreased in all 7 cases. Most common epidermal changes seen were hyperkeratosis and acanthosis. Hyperkeratosis was seen in 4 cases, acanthosis was seen in 3 cases while 2 cases had parakeratosis. Number of mononuclear cells were increased in 2 cases while one case showed presence of mast cells in the dermis. The various clinical and histological features of these cases studied are summarized in table 1.

DISCUSSIONS

Nevus lipomatosus cutaneous superficialis is a rare hamartomatous benign condition characterised by presence of mature adipose tissue ectopically in the dermis. NLCS is more commonly present from birth, but can appear later in life as well[3]. However in our study we found this lesion predominantly in adults (71.43%) while only 2 (28.57%) cases were seen in early part of life and those had the lesion since birth. Moreover patients with NLCS, being asymptomatic, presents invariably late to the clinician, in some cases decades after the appearance of the lesion. Its precise pathogenesis is unknown. A proposed pathogenesis is that the precursor cells around dermal blood vessels give rise to mature fat cells[4]. NLCS is differentiated clinically into two forms though they are very much same on histological examination. The Hoffmann Zurhelle form or the classical form is characterised by multiple soft, non tender nodules, which may coalesce to form plaques. This form occurs more commonly in the gluteal and lower back regions and these lesions are present since birth or develop in first few decades of life[5,6]. The solitary form presents commonly as a single pedunculated or dome-shaped nodule[5,6]. Although the solitary form is predominantly found on the buttocks and thighs, it may occur at unusual sites like scalp, axilla, knee, ear and eyelids[6,7]. We found the solitary form as the more commoner form with 5 cases (71.43%) and the classical form as less common form with 2 cases (28.57%). Jones *et al.*[8] as well as Triki *et al.*[9] in their studies found out the solitary form as the most common form, in a series of patients, similar to our findings. Clinically the differential diagnosis may include neurofibroma, papilloma, nevus sebaceous, hemangioma, fibroepithelial polyps, angiolipoma and focal dermal hypoplasia[10]. In our series we had provisional clinical diagnosis of these lesions as fibroepithelial polyps, papillomas, neurofibroma and soft tissue tumours but none as NLCS. Thus histopathological examination of these lesions solve the puzzle and give the final diagnosis and help the clinician come to the terms of clinical presentation of NLCS. The differential diagnosis of NLCS histopathologically are Goltz's syndrome which has mature fat within the superficial dermis but can be differentiated by conspicuous absence of collagen, clinical appearance, associated underlying deformities and the X-linked dominant inheritance. Melanocytic nevi may shows adipocytes in dermis but they have nevus cell nests in dermis or epidermis or both, and the amount of adipocytes mostly is less than 20%. Large skin tags or lipofibromas which may show adipose tissue in the dermis and are in the same location as nevus lipomatosus. The prominent pedunculated appearance and usually a later onset in life help to distinguish it from nevus lipomatosus[10]. Focal dermal hypoplasia show loosely arranged collagen fibers in the papillary dermis and the adipose tissue is continuous with the subcutaneous fat and extends almost to the undersurface of the epidermis in some areas. Extreme attenuation of collagen in focal dermal hypoplasia helps in distinguishing it from NLCS. These hamartomas mostly have invariably asymptomatic course, although it may in rare instances have unusual growth or morphology such as giant NLCS, comedo-like plugs and foul-smelling discharge,[11]

Table 1: Clinicopathological features of NLS

Sex	Age (years)	Site	Initial clinical diagnosis	Clinical form	Histological features
Male	53	Thigh	Papilloma?/skin tag?	Solitary form	Adipocytes indermis, increased collagenous tissue, Hyperkeratosis, acanthosis, parakeratosis
Female	3	Gluteal region	Fibroepithelial polyp	Classical form	Adipocytes in dermis, Hyperkeratosis, acanthosis, increased collagen in dermis
Female	27	Thigh	Lipoma	Solitary form	Adipocytes in dermis, increased collagenous tissuein dermis, mast cells in dermis.
Female	65	Axillary region	lipoma	Solitary form	Adipocytes in dermis, hyperkeratosis, increased collagenous tissue, increased vascularity in dermis
Male	2	Lower back	Soft tissue tumour?	Classical form	Adipocytes in dermis Hyperkeratosis, acanthosis, parakeratosis, increased vascularity in dermis
Male	17	External auditory canal		Papilloma	Solitary formAdipocytes in dermis, increased collagenous tissue, chronic inflammatory cell infiltration of dermis.
Famale	38	Arm	Neurofibroma	Solitary form	Adipocytes in dermis, increased vascularity, lymphocytic infiltration.

M: male F: female

and ulcerated lesions after external trauma or ischemia[12]. Recurrence have been rarely reported and malignant transformation has never been reported[13]. In our study no recurrence of any case of NLCS was reported till the finalization of this study. This study helped to summarize the various histological features of this rare hamartoma.

CONCLUSION

This is the first study about NLCS in this part of world and summarizes the clinical and the histopathological features of NLCS. Most of the studies about NLCS have been limited to case reports and any case series is a welcome process for imparting knowledge about this hamartoma. In our region solitary form of the NLCS has been found to be the commonest form and most of the cases were seen in adults unlike the general consensus. None of the cases in our study had NLCS as an initial clinical diagnosis and as such clinicians weren't aware of this hamartoma beforehand and as such this study can make the clinicians familiar with this condition so that they can plan the treatment accordingly.

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