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CLINICAL AND HISTOPATHOLOGICAL ANALYSIS OF NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS 9 CASES

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ABSTRACT

Background: A hamartoma is mostly a benign mass of disorganized tissue native to a particular anatomical location. Most hamartomas are usually benign. Nevus lipomatosus cutaneous superficialis (NLCS) is a rare hamartoma of mature adipose tissue. **Aim and Objective:** To study the clinical and histopathological features of 9 patients with NLCS. **Method:** A retrospective analysis of clinical data and histopathological findings in 9 cases of NLCS in our Hospital between 2017 to 2021. Demographic and clinical data were obtained from the clinical case files. Slides and tissue blocks were retrieved. H&E and Masson's trichrome and reticulin stained slides were studied in all cases. **Results:** We identified 9 patients with NLCS (solitary 7 and classical 2) in this series, of whom 5 were males and 4 females. The average age at diagnosis was 53.4 years. The most common location was the lower part of the body. Most of the cases were diagnosed as skin papilloma before skin biopsy. Surgical excision was effective in all patients and on follow up no recurrence was observed. **Conclusion:** Awareness of NLCS among Clinicians is important .Histopathology is confirmatory for diagnosis.

KEYWORDS : Nevus lipomatosus cutaneous superficialis, Skin tumor, Histopathology

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign idiopathic hamartomatous anomaly of the skin. It was first described by Hoffmann and Zurhelle in 1921 as pedunculated lipofibroma. Histologically it is characterized by the presence of clusters of mature ectopic adipocytes amongst dermal collagen bundles without connection to underlying subcutaneous fat.[1,2]

NLCS most commonly presents in birth or infancy as a developmental anomaly (nevus angiolipomatosis of Howell), but it can appear later in life [3]. There is neither sex predilection nor hereditary predisposition in this disorder and is frequently devoid of any associated congenital defects.[2,3]

Clinically two patterns of NLCS exist—classical type (multiple form) and solitary form (rare form). Classical type lesions are the most common form. They commonly occur at the pelvic girdle, lower trunk, gluteal region, and thigh as multiple skin-colored or yellowish papules, coalescing to form plaques with zosteriform, linear, or segmental distribution. These lesions are congenital and usually present at birth or within the first 3 decades of life. They are slow-growing, and if untreated, can attain large size.[1,2]

Solitary form usually presents late onset during the third to sixth decades of life as a solitary dome-shaped sessile papule or nodule, mimicking skin tag. There is no specific distribution including unusual sites like the scalp, eyelid, nose, and clitoris.[1-4]

Generally, there are no clinical symptoms during its course, although sometimes NLCS can appear with some symptoms. Here we report a series of 9 cases of NLCS (initially misdiagnosed as skin papilloma or skin tags) in order to increase awareness about this rare disease and to emphasize the importance of its histopathology.

METHODS

We retrospectively reviewed the records and histopathology of 9 patients with NLCS. They were seen in Surgery or Dermatology OPDs of tertiary health multispecialty hospital between 2017 to 2021.

The following characteristics were recorded for analysis: age, gender, clinical features, histopathology, and treatment. All the routine laboratory investigations and Viral markers (HBsAg/ HCV/ HIV) were performed and the excision of the lesions was done under local anesthesia as daycare surgery and sent for histopathological examination. H&E and Masson's trichrome and reticulin stained slides were studied in all cases. All histopathology reports were reviewed by the Pathologist.

Cases Presentation

Case 1:
A 56-year-old male patient showed the two plaques in the gluteal cleft and back, measuring 1.5×0.9×0.5cm and 1×0.6×0.4cm respectively. Clinically suspected Papilloma.

Case 2:
A 43-year-old female patient had growth measuring 2×2×1.5cm on the right upper chest 5 cm below the clavicle, clinically diagnosed as skin tag .

Case 3:
A 78-year-old male patient had a single growth measuring 4 x3x1 cm on the right inner thigh. Clinically suspected Papilloma.

Case 4:
A 63-year-old male patient presented with a single growth measuring 3.7x2.7x1.5 cm on the lower part of the left thigh. Surface ulceration was seen.Clinically suspected Papilloma.

Case 5:
A 52-year-old female patient had a single growth measuring 2x1.2x0.5 cm on the posterior aspect of the right thigh. Clinically diagnosed as Wart.

Case 6:
A 22-year-old male patient had two growths over the right lower back and right flank, measuring 3x2x1.5 cm and 2.5x1.8x1 cm respectively. Clinically Suspected Neurofibroma

Case 7:
A 57-year-old female patient, Physical examination showed a single polyp measuring 6.5x5x4 cm over the left calf. Clinically suspected Papilloma.

Case 8:
A 53-year-old male patient was clinically diagnosed with a single plaque measuring 2.5x2x0.5 cm over the left scapula (back).Clinically suspected Papilloma.

Case 9:
A 57-year-old female patient's physical examination showed a single growth measuring 2x1x1.2 cm over back. Clinical suspicion was Wart Surgical excision of the lesion was performed in all patients.

RESULTS

Seven cases of NLCS were identified between 2017 to 2021. Five of the nine cases were male. The mean age at diagnosis was 53.4 years. Out of nine cases, five cases were initially clinically diagnosed as skin papillomas, three cases as a wart, and one as neurofibroma. Only one patient had the lesion since birth while the other eight patients had acquired the lesion through the third to eighth decades.

The type was solitary in seven patients and classical in two patients. All patients underwent surgical excision of the lesion and tissue was sent to the pathology lab for diagnosis. On gross, the lesion was globular and the cut surface was homogenous yellow (Fig. 1a &b). Microscopy revealed a polypoid skin lesion. High power view shows unencapsulated lobules of ectopic benign mature adipose tissue and strands of collagen fibers in dermis. There was no connection with underlying subcutaneous tissue. Skin adnexal structures noted. Mild lymphocytic infiltrate seen in the dermis. Masson's trichrome staining was performed for collagen fibers. (Fig. 2)

DISCUSSION

Histopathological evaluation is required for diagnosis of NLCS .Many clinical dermatoses may have a similar clinical presentation such as papillomas (skin tags), plexiform neurofibroma, nevus sebaceous, connective tissue nevus, vascular malformation, and lipoblastomatosis[2,3,4] .Similarly in this study eight cases were clinically diagnosed as papilloma/wart .One had surface ulceration. Ulceration over growth can be worrisome feature to clinicians and patients as well.

On Histopathology NLCS should be differentiated from focal dermal hypoplasia (Goltz syndrome), lipofibromas, and giant acrochordons with fat herniation. Focal dermal hypoplasia/ Goltz syndrome is associated with congenital defects with the presence of fat cells along with the absence/extreme attenuation of collagen in the dermis.[3,4,5]Lipofibromas contain adipocytes, but dermal skin appendages are absent. Giant acrochordons (skin tags) show absence of fat cells in the dermis [1,3].Rare features like increased basal pigmentation and focal elongation of rete pegs, reduced adnexal structures, and abnormal folliculosebaceous structures .[2,6, 7]. These features are not present in our study.

The pathogenesis of NLCS is still not well defined and various theories have been proposed. Buch AC et al suggested that the deposition of ectopic adipocytes is caused by degenerative changes in the dermal collagen bundles and elastic tissue or may originate from precursor cells of the dermal vessels or from pericytes, as in fetal lipogenesis [6,8].

Most published studies describe patients with the classical form of NLCS.[2,9]Only one study describes a preponderance of the solitary type to date[10]. In our series, most of the lesions (7 out of 9) were of solitary type. The onset was late (third to the eighth decade). Interestingly in our series, most of the lesions were over the lower limbs and on back. The size of the lesion varied from 1×0.6×0.4 cm to largest measuring 6.5x5x4 cm. Rarely, huge giant forms of NLCS have also been reported measuring up to 40x28cm [11,12,13].The cut surface was homogeneously yellow and of softer consistency in all cases. This is in contrast to a papilloma which is firmer and more whitish.

NLCS have an asymptomatic and static course. Some lesions may have unusual growth or morphology such as giant NLCS [11,13], comedo-like plug [14], foul-smelling discharge [1,14], and ulcerated lesions after external trauma or ischemia [2, 3]. Recurrence has rarely been reported [1].There are no systemic abnormalities or malignant transformation [5,11]. If left untreated NLCS can increase in size. Treatment is not necessary unless for cosmetic reasons. Wide local excision is first choice ; cryotherapy can also be used.

CONCLUSION:

This study highlights need for awareness of NLCS as correct preoperative clinical diagnosis was not present in any case. The larger lesions were diagnosed as papilloma whereas the smaller ones as a wart. Histopathological examination is mandatory for a final diagnosis of NLCS. Solitary type of NLCS was more common in adult females and the thigh appeared to be the commonest site.

Figures:

Fig 1a& 1b: NLCS, polypoidal cerebriform growth and yellowish-white cut surface.

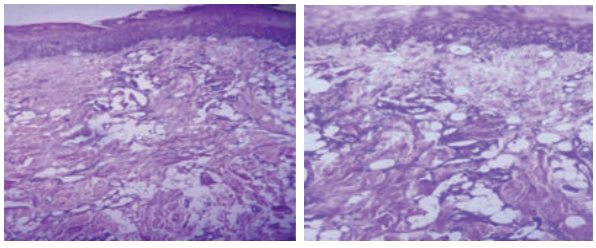

Fig 2: a) Low power view (X10) of NLCS showing the presence of ectopic mature adipocytes in the superficial dermis and disorganized collagen bundles in the dermis.

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