Nevus Lipomatosus Cutaneous Superficialis: Clinico-Pathological Study from a Tertiary Center in Nigeria

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ABSTRACT

Nevus Lipomatosis Cutaneous Superficialis (NLSC) is a rare benign hamartomatous skin disorder. It may present in a classical multiple cerebriform nodular form or rare solitary type. It is defined, histologically, by dermal deposition of Adipocytes. We report herein a case series of six patients who had predominantly, classical presentation.

Keywords: nevus lipomatosus cutaneous superficialis, Tertiary center, Nigeria

Declaration of conflict of interest: We declare no conflict of interest

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare benign hamartoma characterized by the presence of mature adipocytes in the papillary dermis. It presents in two forms, multiple classical and rare solitary varieties. The latter appearing in adult life, while the former may be present at birth or in the first two decades of life. We report here 6 cases of Nevus lipomatosus cutaneous superficialis some of which are on unusual sites.

METHOD

A 10-year retro respective case series was conducted between January 2005 and December 2014 involving all patients with histologically confirmed NLCS at the dermatology clinic of Aminu Kano Teaching Hospital Kano.

RESULTS

A total of six patients were documented. The epidemio-clinical characteristics is summarized in Table 1. Of the six patients, three were males. They are all aged between 9 and 55 years. Two had the disorder since childhood; it appeared in another two in the second decade of life, and in the last two, thereafter. In one case the appearance of the lesion was preceded, at the same spot, by a café-au-lait patch at birth. In all the 6 patients, NLCS presented as asymptomatic, slowly progressive soft lesion over years. No association with other cutaneous disorders was seen in all cases. In addition, no systemic symptoms were reported. Classical form of NLCS was observed in four patients while the other two patients had solitary form (Figures 2&3). The lesions involved the limbs in four patients and the shoulders in two others. Systemic examinations were not remarkable in all the patients. Routine laboratory examinations were all normal.

Biopsy of the lesion in all the six cases revealed aggregates of mature adipocytes in reticular dermis. In 3 cases the fatty cell extended into the papillary dermis (Figure 4). None of the specimen showed linkage of these adipocytes with the underlying subcutaneous fat.

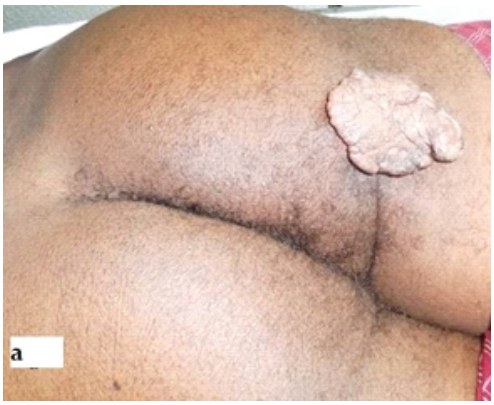
Five patients underwent surgical excision, following successful referral to plastic surgery unit and one case declined treatment (Figure 2).

DISCUSSION

It was in 1921 that Hoffman and Zuhrelle first described Nevus lipomatosis cutaneous superficialis as a rare hamartomatous benign condition of unknown cause.3 The condition exists in two forms, the common classical and the rare solitary type. The classical form presents as a group of multiple, non-tender soft cerebriform nodules. The nodules may coalesce to form unilateral plaque in a segmental,

S/N	Age	Sex	Duration	Location	Clinical Aspect
1.	55	M	4	Right buttock	Pedunculated plaques
2.	24	F	18	Right shoulder	Soft skin-colored 2-3cm grouped nodules
3.	18	F	5	Left shoulder	Multiple soft nodular cerebriform tumour
4.	9	F	4	Right sole	Solitary dome-shaped yellowish mass
5.	20	M	6	Left popliteal fossa	Skin-colored, soft, cerebriform plaque
6.	32	M	8	Right thumb	Soft dome-shaped yellowish nodule

Table 1. Epidemio-clinical characteristics of the patients with NLCS



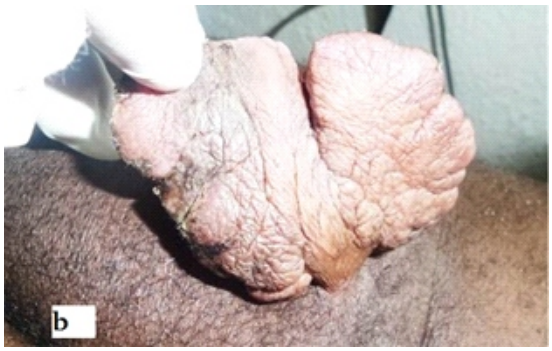
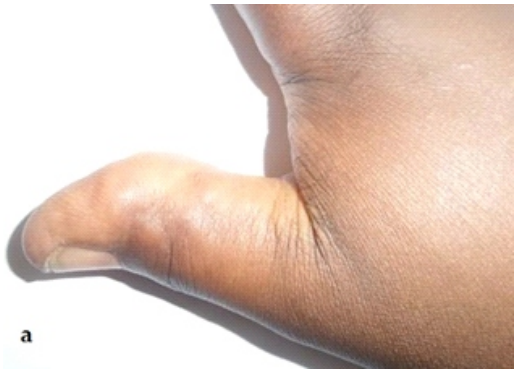


Fig. 1 (a&b): Soft flesh colored Pedunculated plaque



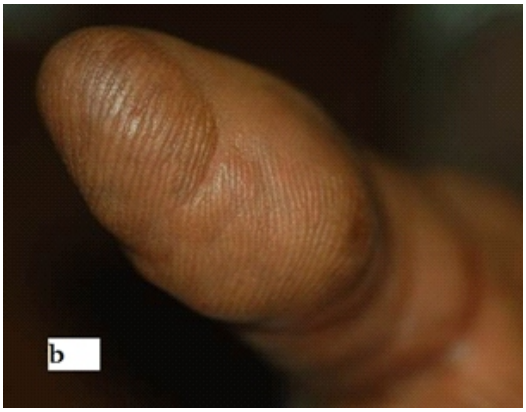


Fig. 2 (a&b): Flesh-colored lobulated cutaneous mass

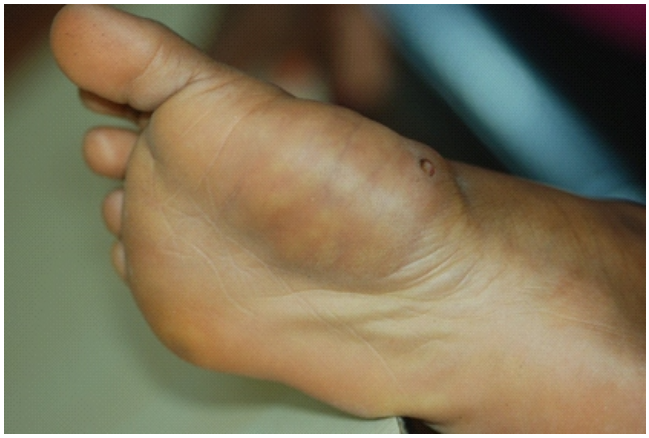


Fig. 3: Yellowish, dome-shaped lobulated cutaneous mass (8cm x 6cm).

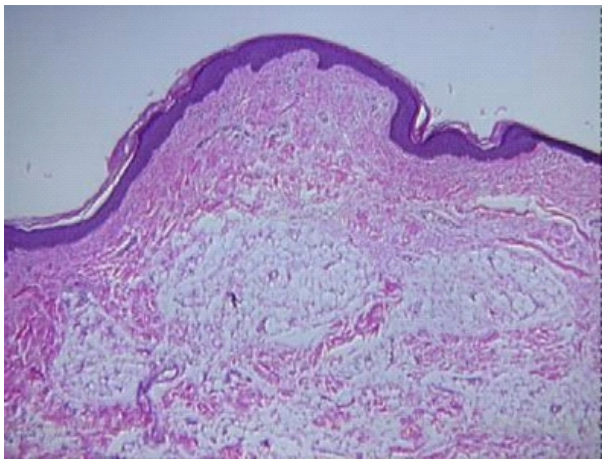


Fig. 4: Groups and strands of mature lipocytes imbedded between collagen bundles in the reticular dermis X10

zosteriform or linear distribution. The lesions are often seen around the pelvic girdle involving the buttocks, sacral or lumbar region. Rarely, the abdomen, chest, face or the scalp may be involved.4, 5, 6

The rare solitary form of NLCS present as a solitary nodule or a pedunculated mass that appears in adult life. In contrast to the Classical form, the solitary type has no predilection site.7 It has been reported on many sites including the nose and clitoris.8, 9

In this series, two of the cases had shoulder presentation. The two other cases involving the thumb and sole of the foot, are to the best of our knowledge never documented. None of the six reported cases had family history. Likewise, there has not been evidence of a familial tendency or sex predilection in earlier reported cases.10 Similar to the six cases we reported, NLCS is almost always asymptomatic, although ulceration following external trauma or ischemia had been reported.11 Coexistence of NLCS with other cutaneous disorders such as café au-lait macules,, leucodermic patches, overlying hypertrichosis, and comedo-like alteration has been reported.1 Café-au-lait patch was noticed on one of the 6 cases in this report. So far, NLCS has not been associated with systemic abnormalities or malignant changes.12 Although so many assumptions have been made, the pathogenesis of NLCS still remains unknown. There is a possible relationship between NLCS and connective tissue nevi, but, further studies are needed for confirmation and clarification. It still remains inconclusive the role of genetic abnormalities in the

development of NLCS. Though, recently, a report of a NLCS with a 2p24 deletion has been reported.13

Clinically, the differential diagnosis of NLCS may include neurofibromatosis, lymphangioma, hemangioma, skin tag or fibroepithelial polyp.14

Histological features in both the two clinical forms is similar. The histology shows groups of adipocytes intermixed with collagen bundles. The adipocytes may extend into the papillary dermis. Characteristically there is no connection between these adipocytes and the adipocytes in the subcutis. A similar dermal collection of adipocytes may be seen in some Melanocytic nevi, pedunculated lipofibromas and in Gotz syndrome. However, the absence of skin appendages in the dermis in both Gotz syndrome and Lipofibromas is a distinguishing factor.

Although excision is curative and, recurrence after surgery is rare, treatment might not be necessary other than for cosmesis17. Five of our patients had surgical excision and, so far none had reported any complications or recurrence. If surgery is not an option, alternative therapies include cryotherapy18 or CO2 laser.19

CONCLUSION

NLCS is a rare benign idiopathic skin malformation; though not known for malignant degeneration ; early recognition and resection eliminates the need for extensive reconstruction of the defect and post-operative scar formation.

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