

Nevus Lipomatosus Cutaneous Superficialis: Poses a Wholistic Health Problem: A Case Report

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Abstract: *Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare benign developmental anomaly characterized by accumulation of mature fat cells within the upper dermis. Two forms were described and the classical multiple forms were first reported by Hoffman and Zurhelle in 1921. The other being the solitary form. These two forms may occur at birth or may appear at later age. There is no specific gender predilection. Here we report a case of Giant classical type NLCS in a 20-year-old woman.*

Keywords: *Nevus Lipomatosus Cutaneous Superficialis, benign fat tumor, surgical excision*

I. Introduction

Nevus Lipomatosus Cutaneous Superficialis is a rare idiopathic dermatosis¹ characterized by isolation of mature adipocytes within the dermis.^{2,3} Two clinical variants were present of which the classical form was reported first by Hoffman and Zurhelle⁴ that usually present from birth. The solitary form is less common and develops at a later age as a solitary papule or nodule.^{5,6} In a large retrospective case series various skin abnormalities such as follicular papules, hypertrophicpilo-sebaceous units, angiokeratoma of Fordyce, leucoderma macules, and hemangiomas have been shown to be associated with NLCS.¹ Dhar S et al reported ulceration of the nevus. Few conditions having similar features of NLCS such as xanthoma, nevus sebaceous, plexiformneurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (Goltz syndrome) should be considered under differential diagnosis.⁸ Treatment is not required except for cosmetic reasons when surgical excision is the treatment of choice. Here we report a case of Giant Hoffman-Zurhelle type of NLCS located over the lumbar area causing socio-sexual as well as cosmetic problem in a 20-year-old woman.

II. Case Report

A 20-year-old woman attended outpatient Department of Dermatology, Andhra Medical College (Visakhapatnam, Andhra Pradesh, India), with the complaint of large asymptomatic mass over the lower back since birth. The lesion started as small multiple papules and nodules that progressively increased in size and attained the present size. No similar lesions were reported among their family members.

On physical examination there was a large mass across the lower back extending on either side of the midline. On dermatological examination a single large mass of 18cm x 15cm in size was seen over L3, L4, and L5 vertebrae extending beyond the vertebral column on both sides. The mass was composed of multiple skin colored soft, non tender nodules of varying sizes that coalesced to form a large plaque having cerebriform surface. Two satellite nodules of 1-2 cm in size were present on the left side of the lesion. The skin over the lesion was non-hairy with normal pigmentation. (Figure: 1)



Figure1: A Large plaque over lumbar area composed of multiple nodules with cerebriform surface.

Examination of the other systems revealed no abnormalities.

A skin biopsy taken from one of the nodules showed collection of mature adipocytes intermingled with collagen bundles in the papillary and upper reticular dermis. The clinical features described by Hoffman and Zurhelle were present in this patient and the histological features were consistent with the diagnosis of nevus lipomatosus cutaneous superficialis. (Figure:2)

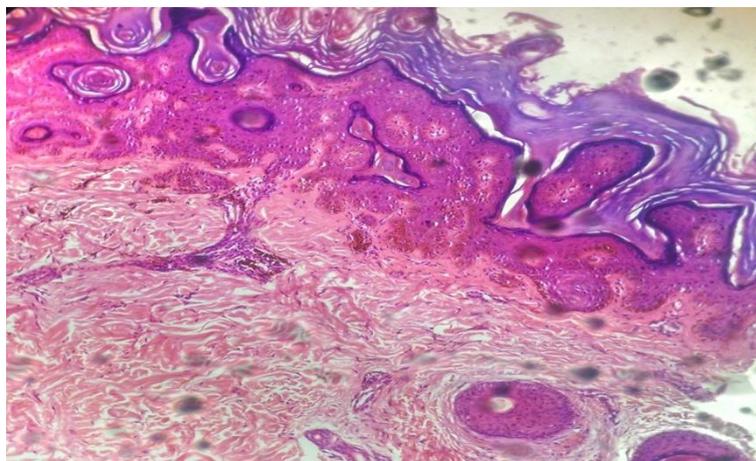


Figure 2: Normal epidermis with mature fat cells in reticular dermis

The patient was referred to the Department of Plastic surgery where the mass was excised and sutured with no post operative complications.

The patient was under follow up with no recurrences since six months.

III. Discussion

NLCS is a rare benign hamartomatous skin lesion.¹ The features described by Hoffman and Zurhelle such as multiple soft skin coloured, cerebriform papules and nodules coalesce into plaques were seen in our case.^{1, 2, 7} Two clinical forms were reported such as the classical multiple forms and the solitary form. No gender predilection⁴ was observed in the literature; however most of the patients were females including our patient. This nevus, the classical form, was characterized by multiple non-tender, skin colored, cerebriform papules and nodules.^{1, 2, 4, 6, 6} Various sites have been reported such as around the pelvic girdle^{1, 2} and several other rare sites.^{5, 10} The solitary form presents as a papule or nodule.² and it occurs in adult life⁴ with no specific gender or site predilection.¹

Other features such as cafe-au-lait spots, hypo-pigmented or leukodermic macules^{5, 6, 9, 10} and ulcerations⁵ over the lesion have been reported which were not observed in our patient.

Under differential diagnosis conditions such as xanthoma, nevus sebaceous, plexiform neurofibroma,¹⁰ lymphangioma, hemangioma, and focal dermal hypoplasia (Goltz syndrome)^{1, 2, 4, 6, 8} should be considered where histology helps in the differentiation of these conditions.

A rare feature such as extension of the lesion beyond the midline described in the literature was present in our patient.

Etiopathogenesis of NLCS is unknown,^{6, 10} but several other postulates were proposed such as degenerative changes or adipose metaplasia of dermal collagen and elastic tissue as the cause for the deposition of the adipose tissue within the dermis.^{1, 2, 5, 6, 8}

Previous reports showed treatment is not necessary⁶ except for cosmetic purpose, and another author showed ulceration¹ over the lesion as an indication for removal of the nevus.

Hence we report a case of giant NLCS located over the lower back causing difficulty during sleep and during delivery. In view of above physical symptoms as well as cosmetic and socio sexual problems, the patient was referred to the department of plastic surgery for surgical excision,

In conclusion we recommend early diagnosis and early excision eliminates extensive reconstruction of the defect as well as reduces post-operative scar formation with good aesthetic appearance.

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