



Naevus Lipomatosus Cutaneous Superficialis of Hoffman and Zurhelle- A Rare Case Report

KEYWORDS

naevus, lipomatosus, hoffman, zurhelle, hamartoma, adipocytes, band-like

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ABSTRACT

Naevus lipomatosus cutaneous superficialis is a rare benign, idiopathic hamartoma of the skin characterized by the presence of clusters of mature adipocytes in the dermis. This is classified into a classical form and a solitary form. The condition was first described by Hoffman and Zurhelle. We report here a case of giant Naevus lipomatosus cutaneous superficialis on the lower back in a bilateral band-like distribution in a 20 year old female.

INTRODUCTION:

Naevus Lipomatosus Cutaneous Superficialis of Hoffmann and Zurhelle is a uncommon cutaneous hamartoma presenting as skin coloured or yellowish soft papules, plaques, nodules or cerebriform masses [1]. There are two clinical forms: (a) the Classical form and (b) the Solitary form. In the classical form, there are multiple papules and nodules in a zonal or segmental distribution. The lesions are seen at birth or develop in the first two decades of life [2]. They are usually seen on the lower back, buttocks and upper thighs. The solitary form can develop in adults and shows a wider distribution in the skin. There is no predominant gender predilection or familial tendency. The lesions are histologically composed of aggregates of mature fat cells interspersed among collagen bundles in the dermis. Usually the number of adnexal structures is reduced as compared to normal adjacent skin but their morphology is not altered. However some cases of naevus lipomatosus cutaneous superficialis with pilar anomalies have been reported such as abortive hair germ-like structures, hypertrophic pilosebaceous units, perifollicular fibrosis and folliculo-sebaceous cystic hamartomas.

CASE REPORT:

A 20 year old female presented with asymptomatic nodular lesions over the lower back of 5 years duration. The lesions were insidious in onset, slowly progressive and gradually increased in size to attain the present size. The lesions were not associated with itching or pain. There was no history of similar lesions elsewhere on the body. There was no history of any treatment taken for the lesions.

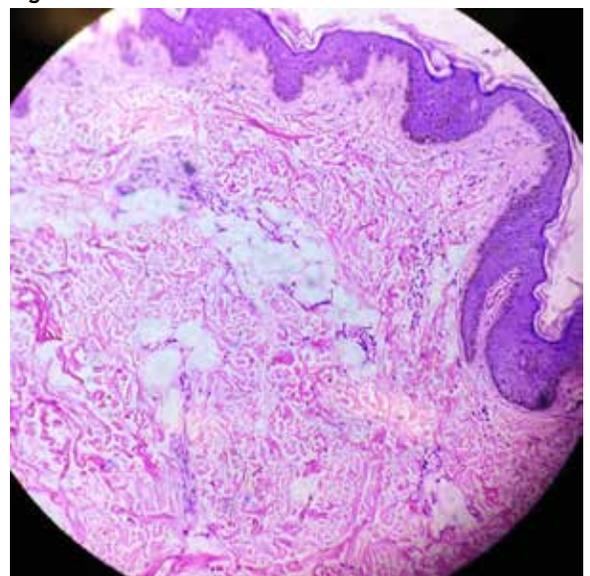
On examination multiple, soft to firm, clustered, skin-coloured, papulo-nodules with cerebriform surface were seen over the lumbar region in a bilateral band-like distribution [Figure-1]. The lesions were non-tender. The overlying and surrounding skin was normal. There were no similar lesions elsewhere in the body. General and systemic examination of the patient were normal. Routine blood investigations, urine analysis and lipid profile were found to be normal. Based on the clinical picture, differential diagnosis of naevus lipomatosus and plexiform neurofibroma were considered. Excision biopsy of a nodule was done and sent for histopathological examination. Histopathology showed mature fat cells in the dermis interspersed among collagen bundles [Figure-2]. A diagnosis of Naevus Lipomatosus Cu-

taneous Superficialis was made. The patient was referred for wide excision for cosmetic reasons.

Figure 1 - Numerous plaques and nodules over the lumbar region



Figure 2 - Clusters of mature fat cells in the dermis



DISCUSSION:

Naevus Lipomatosus Cutaneous Superficialis was described by Hoffmann and Zurhelle in 1921. It is a developmental anomaly which may be present at birth or develop during the first two decades of life. The classical type is frequently seen over the pelvic girdle, buttocks, lower back and upper thighs. This is characterized by plaques or cerebriform masses consisting of papules and nodules in a zonal or segmental distribution [2]. The lesions are slow growing but can reach a large size if left untreated as in our patient. The other type is the solitary form which is seen in adults during 3rd - 6th decades and can occur anywhere on the skin. The histogenesis of naevus lipomatosus cutaneous superficialis remains unknown. It has been postulated that naevus lipomatosus may develop due to fat deposition secondary to degenerative changes in the connective tissue or the fat cells may represent a true naevus resulting from focal heterotropic development of adipose tissue [3]. Epidermis may show basket weave hyperkeratosis, acanthosis, obliteration of rete ridges with focal elongation and increased basal pigmentation. The proportion of fatty tissue in the dermis varies greatly from 10% to 50% [4]. The skin conditions which have been reported in association with naevus lipomatosus are café-au-lait macules, hypertrichosis over the naevus, comedo-like lesions and Angiokeratoma of Fordyce. Naevus lipomatosus should be differentiated from naevus sebaceous, neurofibroma, lymphangioma, cylindroma and angiolipoma [5]. Treatment may be needed for cosmetic reasons. Surgical excision is the treatment of choice and recurrence is rare.

CONCLUSION:

We have reported this case of classical Naevus Lipomatosus Cutaneous Superficialis for its giant size, onset after 15 years of age and the bilateral distribution which is very rare.

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