NAEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS
(Case Report)

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Summary

A case of naevus lipomatosus cutaneous superficialis on the left sacro-gluteo-coxal area in a 25 year old male is reported because of its rarity. The clinical features and histopathological changes of this condition are described. The possible factors regarding its predilection to the pelvic girdle region, the various theories regarding its pathogensis and its differentition from focal dermal hypoplasia syndrome and grouped lesions of macular atrophy are discussed.

KEY WORDS: Naevus Lipomatosus Cutaneous Superficialis, Nevoid disorder, ectopic fat cells, Focal dermal hypoplasia syndrome, hetertrophic development, Lipophilic.

Introduction and literature review

Hoffmann and Zurhelle, in 1921, first described the clinical condition called "naevus lipomatosus cutaneous superficialis!" This condition is a rare nevoid variety of lipoma of the skin[2] and is clinically and histopathologically a distinct entity. The lesion is usually evident at birth and may increase with age[3]. In some cases lesions have appeared much later and in one case described by Holtz (1955), the naevus appeared at the age of 18 years[4]. It has been regarded as a developmental anomaly, since it has often been noted at birth[5]. Common sites of occurrence are the gluteal region, low back, sacral and coccygeal regions[6], but lesions have been described over the upper thighs[6], the ear, the scalp, the abdominal wall, shoulder, legs and ankles[5]. Some of the reasons offered for the frequency of these lesions in the pelvic girdle region are peculiarity of circulation in this region, persistent pressure during intrauterine life (Nikolowski, 1950)[7] and perhaps a 'lipophilic' nature of the fat pad here which is usually thick and frequently also a site of predilection for certain other disorders of fat such as lipodystrophies and dystrophia adiposogenitalis[4]. No predilection to any sex has been observed.

Two clinical variants of naevus lipomatosus cutaneous superficialis are described, the large plaque type and the solitary nodular type. In the former

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the plaques consist of aggregations of non-tender, skin coloured or pale yellow papules and nodules, usually smooth but at times irregularly folded or even warty. They are soft in consistency. The plaques vary in size reaching even upto 8 – 15 cms. The lesions may be localised, linear or systematized[6]. No other associated abnormalities have been described so far.

The origin of this unusual disorder remains obscure. Various theories have been put forward to explain the pathogenesis :

Based on the presence of degenerate collagen and fragmented elastic tissue Hoffmann and Zurhelle (1921) considered the deposition of fat to be secondary to some degenerative change in connective tissue[1]. Though Nikolowski (1950), agreed with these such changes, have not been seen uniformly. The theory that primary disturbance is in the growth of adipose tissue from blood vessels and the degenerative changes in the connective tissue are secondary was advanced by Haltz (1955) and supported by Lynch-Goltz[8] because both mature fat cells and immature cells resembling embryonic adipose tissue may be present in this condition[4].

Focal heterotopic development of adipose tissue in the corium was proposed by Robinson and Ellis[19] who suggested that the displacement of subcutaneous adipose tissue into the dermis takes place during foetal development. The fact that the dermal fat deposits are not contiguous with the subcutaneous fat in majority of the cases does not entirely negate this possibility. It is now generally agreed that the nevus lipomatosus represents nevoid anomaly in which ectopic fat cells have formed in the dermis from the perivascular mesenchymal tissue[10].

In a pure naevus lipomatosus histology shows groups and strands of mature fat cells embedded among the collagen bundles of the dermis, often upto the subpapillary layer. In the deeper dermis the fat cells surround large blood vessels[5,8]. The larger nodules that may occur in some cases consist for the most part of mature collagen bundles, among which are scattered small groups of fat cells. Serial sections may show connections of the dermal aggregates of fat cells with the underlying subcutaneous fat in some cases and no such connections in other cases[9].

Lynch and Goltz pointed out that the epidermal changes in this nevus generally resemble those of epidermal naevi[8]. Hyperkeratosis of basket-weave type, follicular plugging, moderate acanthosis, squaring of retepegs and increased pigmentation of the basal layer have been described.

The similarity between the clinical and histological findings in all the reported cases suggests that this is a distinct entity. To our knowledge there is only one case reported from India by Shroff et al[10]. We are reporting the case because of its rarity.

Case Report

A 25 year old male agriculturist attended the outpatient department of Skin & S.T.D., J.J. M. Medical College with symptomatic skin lesions over the left sacral region present since birth. At birth his mother noticed a small soft skin lesion over the left sacral region and since then this lesion had increased to the presenting size.

Examination revealed multiple round papules, nodules and plaques of varying size and shape situated on the left sacro-gluteo-coxal area. The lesions were light brown to pale-yellow, soft non-tender and non-adherent to deeper structures. Surface was smooth in

some areas and lobulated at other areas and the total area of involvement was 8 × 5 Cms. (Fig. 1). Systemic examination was normal and the patient's general health was good. There was no evidence of any other associated nevoid disorder.

topathological features were consistent with those of naevus lipomatosus cutaneous superficialis.

Discussion

Nevus lipomatosus cutaneus superficialis is a rare condition with dis-

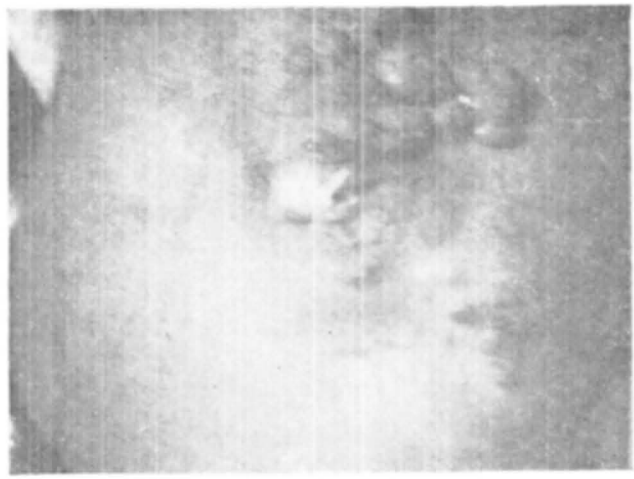

Fig. 1

Shows Naevus Lipomatosus cutaneus superficialis on the left Sacro-gluteo coxal area.

A provisional diagnosis of a connective tissue naevus was made and a biopsy was taken. Histopathological sections showed papillomatosis and mild hyperkeratosis (Fig. 2). There is dense collagenization of the dermis which showed focal aggregates of mature fat cells in the upper dermis (Fig. 3). In the deeper dermis fat cells were seen around blood vessels. Histinctive features to make it an entity Our case conforms well to this rare condition both clinically and histopathologically. Lesions that may resemble nevus lipomatosus cutaneous superficialis occur in focal dermal hypoplasia syndrome. This syndrome has derived its name from one of its most important features, namely, undeveloped skin through which thin,

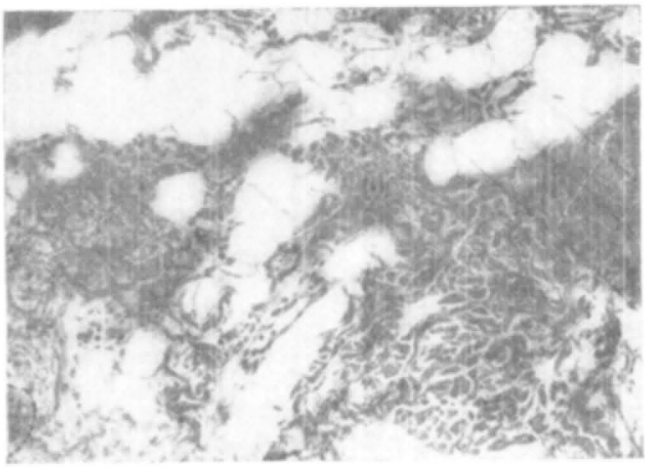

Fig. 2

Shows basket-weave type hyperkeratosis (mild), keratotic plugging, squaring of retepegs and focal aggregates of mature fat cells in the deeper dermis (× 100)

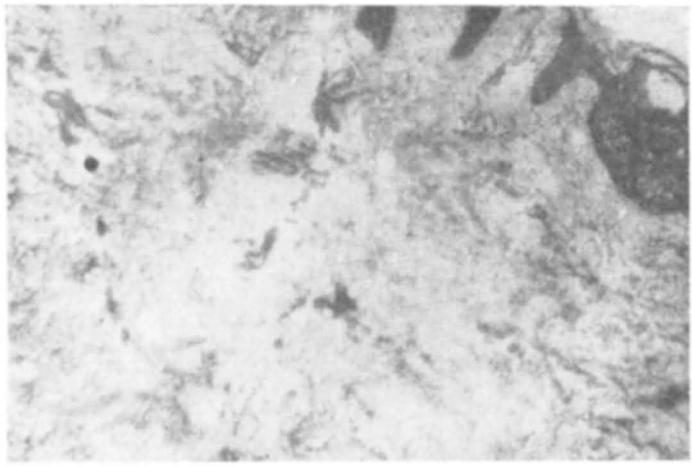

Fig. 3

Groups and strands of mature fat cells are embedded among the collagen bundles of the dermis extending as high as the subpapillary layer. Portions of the epidermis are seen at the top (× 100)

subcutaneous fat herniates. In focal dermal hypoplasia syndrome multiple developmental abnormalities of both ectodermal and mesodermal structures have been described involving skeletal, ocular, oral, dental and soft tissues[5]. There are histological similarities between focol dermal hypoplasia and nevus lipomatosus[12], but there is an extreme degree of attenuation of collagen in focal dermal hypoplasia. Grouped lesions of macular atrophy may be confused with nevus lipomatosus cutaneous superficialis, but a careful history, clinical examination and histopathological features will easily differentiate the two conditions.

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