Solitary nevus lipomatosus cutaneous superficialis

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A 7-year-old female presented with a swelling over the right calf since childhood. The swelling increased in size to the present size following trauma. On examination she had a soft, slightly tender swelling measuring 3 x 2.5 cm over the right calf. The skin over the swelling was dark, thickened and wrinkled. Clinical diagnosis was infected hematoma. No similar lesions were noticed elsewhere. There was no family history of similar lesions. Excision biopsy was performed and submitted for histopathological study.

Grossly, the specimen consisted of a dark brown skin-covered mass measuring 3.5 x 2 x 1 cm. The skin over the lesion was wrinkled and thickened (Figure 1). The cut surface was yellowish with few brownish spots. Microscopically the sections revealed epidermis with mild hyperkeratosis, focal acanthosis and follicular plugging. The dermis showed clusters of mature fat cells predominantly in the middle and deep dermis (Figure 2). The papillary and subpapillary dermis showed ectatic capillaries which may be responsible for the dark brown skin colour. Some of the capillaries were cuffed by a variable number of spindle-shaped and round mononuclear cells (Figure 3). A histological diagnosis of nevus lipomatosus cutaneous superficialis was given.

Discussion

Nevus lipomatosus cutaneous superficialis is a rare malformation characterized by ectopic adipose tissue in the dermis. The condition was first described by Hoffman and Zurhelle in 1921.[1] Few cases have been reported thereafter.

The lesion is present mostly since birth or noticed in the first two decades of life, with no sex predilection. Clinically it can be multiple or solitary, soft, skin-coloured papules sometimes coalescing to form plaques with a cerebriform surface. Multiple lesions are seen more commonly in younger patients while...
a solitary lesion is generally seen in those over 20 years of age. It could be that a solitary lesion is ignored in childhood as a non-specific hamartomatous lesion or that those with solitary asymptomatic lesions tend to present later in the twenties for cosmetic reasons. The multiple lesions have a marked predilection for the gluteal region, lower back and upper thigh[1] though solitary lesions may occur at unusual sites like the scalp, clitoris[1,2] and in the calf as in the present case.

The peculiar histopathological finding is the presence of ectopic fat, which may vary from 10-50% of the total lesion, in the dermis.[1] Most of them are mature fat cells and may be present in the perivascular area. All the studies have documented increased vascularity in the subpapillary and papillary dermis with perivascular mononuclear cell and spindle-shaped cell infiltration. We also observed similar cells. There may be irregularity in the collagen bundles and reduction in the amount of elastic tissue towards the surface.[1] Epidermal changes like mild to moderate acanthosis, basket weave hyperkeratosis, obliteration of rete pegs, increased basal pigmentation and focal elongation of rete pegs have been reported.[1,2] Adnexal structures may be unaffected or reduced in numbers and may show perifollicular fibrosis.[1]

This lesion has a similarity to focal dermal hypoplasia, in which there is a broad range of developmental anomalies with involvement of both the ectodermal and mesodermal structures. It may present with localized scleroderma-like appearance.[4] Differentiating a solitary lesion from giant skin tag with fatty herniation is still a difficult task.

Many theories have been postulated regarding the histogenesis of this lesion. Hoffman and Zurhelle postulated that fat deposition in the dermis is secondary to degenerative changes in the connective tissue. Other theories reveal that the fat cells represented a true nevus that resulted from the focal heterotopic development of adipose tissue, which may occur during embryonal development.[1]

Holtz, in 1955, opined that primitive preadipose tissue grows from the mononuclear cells which they believe were differentiating lipoblasts around the capillaries. This was supported by electron microscopic study demonstrating fat cells in close proximity to blood vessels.[1]

This case is a distinct entity as the lesion is solitary, in a child in the calf region unlike other reported cases. The lesion can be classified as a benign lipomatous hamartomatous lesion, probably nevoid in origin. Perivascular mononuclear and spindle cells noticed in the lesion favors the origin of ectopic fat from these perivascular stem cells or spindle cells indistinguishable from fibroblasts or inflammatory cells on H/E stained sections. The physician should be aware of this rare tumour which can become extremely large if untreated.[5]

References