Nevus Lipomatosus Cutaneous Superficialis (Hoffmann-Zurhelle)

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Nevus lipomatosus of Hoffmann-Zurhelle is a rare condition characterized by ectopic accumulation of adipose tissue that occurs predominantly in the pelvic girdle area (1-3). The lesions are asymptomatic and usually either congenital or seen by the third decade of life (4-6).

Case report

A 23-year-old woman was hospitalized for the evaluation of chronic urticaria. On the physical examination of the skin, we noticed firm, skin-colored multiple papules and nodules ranging from 0.2 to 1 cm in diameter in 16× 20 cm area of the left lower back (Fig.1). These lesions had been present for the last five years, and had increased in size. It was asymptomatic. The patient's general condition was good and all the laboratory findings were within normal limits. A skin biopsy specimen was obtained from the lower left back and stained with hematoxylin and eosin with a clinical diagnosis of connective tissue nevus.

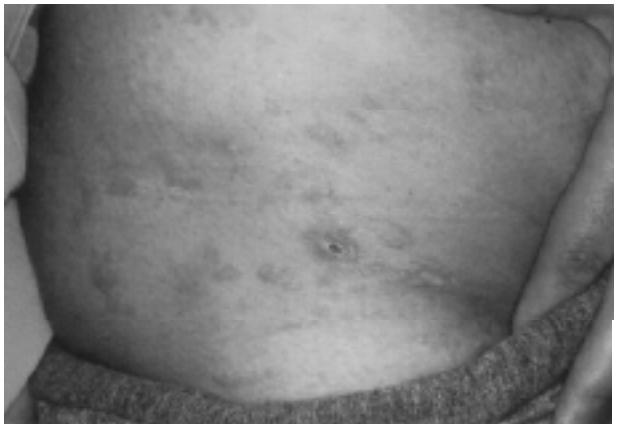

Figure 1. Skin-colored multiple papular and nodular lesions on the left lower back.

Histological examination of the specimen revealed groups and strands of fat cells embedded among the collagen bundles of dermis, some of which being as high as papillary dermis. Densities of the collagen bundles were greater than those in normal skin. The individual fat cells were mature and normal in size. The lobules were not encapsulated and did not communicate with the subcutaneous fat tissue (Fig. 2). Therefore, it was diagnosed as nevus lipomatosus cutaneous superficialis (NLCS). However the patient refused the treatment.

Discussion

NLCS is an uncommon idiopathic skin malformation. It was first described by Hoffmann and Zurhelle as "naevus lipomatodes cutaneous superficialis" in a 25-year-old man with congenital, multiple soft nodules in the left gluteal region (3,5,7,8). Histopathologically it is characterized by groups of ectopic fat cells among the collagen bundles of upper dermis and, it is mainly located on the gluteal region (1-3,5,7).

NLCS is classified into two clinical types, a multiple form (or classic type) and a solitary form (4,7-9). The classic type appears as grouped, yellowish, soft papules or nodules, without tenderness, which often coalesce into plaques on the lower back, buttocks, or thighs in a zonal distribution or along the lines of the skin folds. The lesions usually present at birth but may develop during the first two decades of life. Their surfaces are usually smooth, but rarely they may be wrinkled or verrucoid in appearance (3,7,8). They are varying in size, generally appear at the same time, and usually remain unchanged, once formed, although some continue to enlarge for many years (7-9). A few cases have occurred on the face, scalp, shoulder, thorax, and abdomen (3,7,8). Our patient showed the multiple forms of NLCS on the lower back occurring at the age of 18 with little change in five years.

The second presentation of NLCS is a solitary nodule that usually occurs after the age of 20 years, with no particular distribution. This type of NLCS was met on the ear, scalp, forehead, back, axilla, arm, upper thigh, and knee (5,7,8).

Exact cause of NLCS is unknown and there is no definite explanation why the classic type of NLCS shows such a marked predilection for the pelvic girdle area. Several theories have been proposed to explain the pathogenesis of NLCS (9). Hoffman and Zurhelle thought that deposition of adipose tissue was secondary to degenerative changes in dermal collagen and elastic tissue.

The histological appearance of the clusters of ectopic mature adipocytes among the collagen bundles or around

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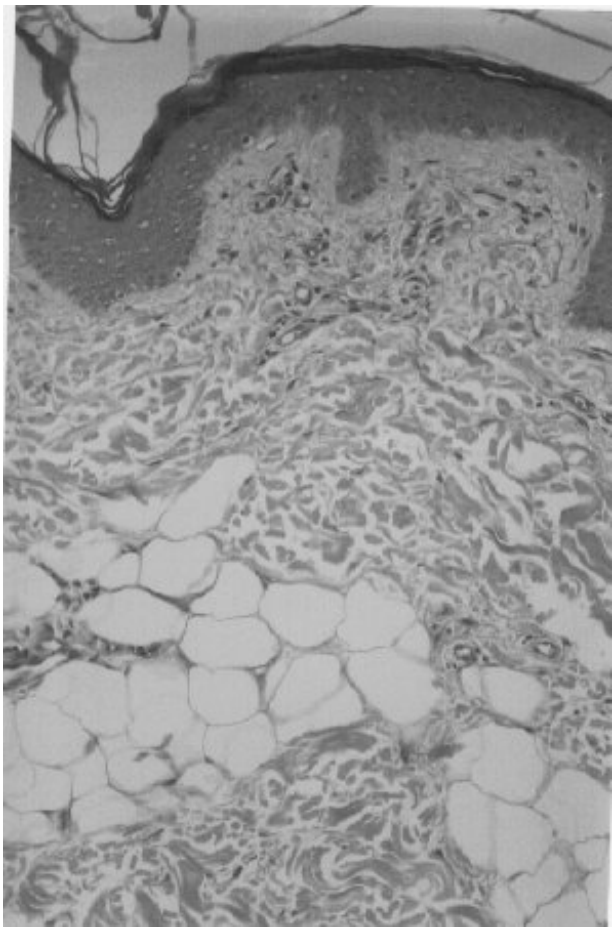

Figure 2. Groups and strands of mature fat cells are embedded among the collagen bundles of the dermis, extending as the subpapillary layer (Haematoxylin and eosin, X25).

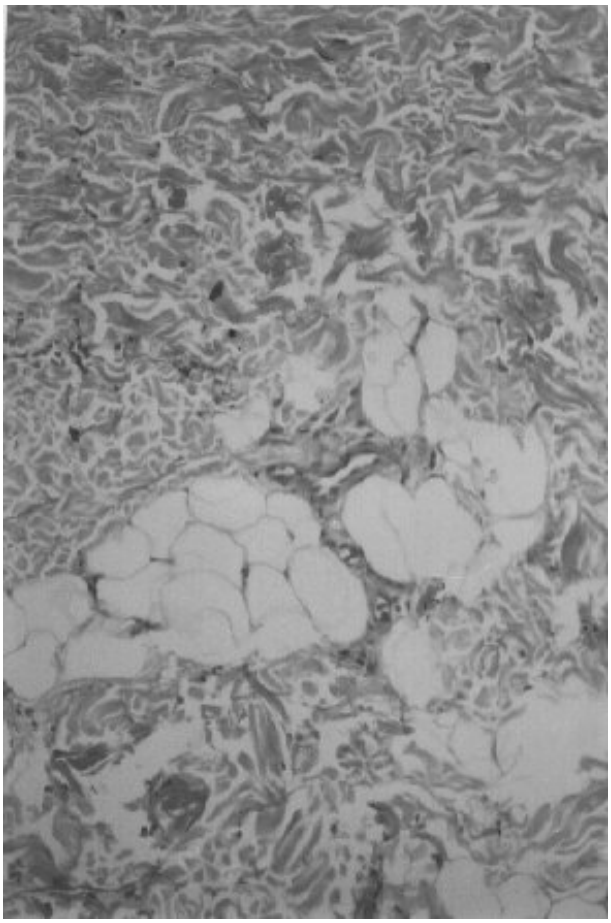

Figure 3. The fat cells are apt to be situated in small foci around the subpapillary vessels (Haematoxylin and eosin, X50).

the blood vessels of the subpapillary plexus and also within the level of the papillary dermis is the most outstanding microscopic feature of NLCS (1,5-7,9). Additional features are abnormality of the epidermis and appendages, increased vascularity, a greater number of fibroblasts, and irregular distribution of collagen and elastic fibers in the dermis. Smaller foci of fat cells are often seen in a perivascular arrangement (1,4,8). A biopsy specimen of our case revealed that groups of ectopic fat cells were found at the level of the papillary dermis and that they were in the vicinity of blood capillaries.

Clinically, the differential diagnosis may include neurofibroma, lymphangioma, haemangioma, and giant fibroepithelial polyp (7,8). Histologically, adipose cells located in the dermis are often seen in focal dermal hypoplasia (10). However, these two entities are clinically different. Focal dermal hypoplasia is associated with several ectodermal and mesodermal deformities including syndactyly, hypoplasia of the hair, nails, and teeth, cutaneous ulcers with thinning of the skin, and herniations of adipose tissue in yellowish papules. Microscopically the dermis is almost entirely replaced by fat cells (8).

Treatment is not warranted and is usually not necessary in NLCS other than for cosmetic reasons, in which simple surgical excision would be enough. This appears to be curative for the lesion does not recur after excision. The lesions change little with time (8). Systemic abnormalities and malignant changes have not been associated with NLCS (6,8).

References

1. Atherton DJ: Naevi and other Developmental Defects. In: (Eds. Champion RH, Burton JL, Burns DA: Breathnach SM) Textbook of dermatology, 6th ed. : Blackwell Science Ltd, Oxford; London 1998, pp: 519-616.
2. Lever's Histopathology of the Skin [Text and Atlas on CD-ROM] Lippincott Williams & Wilkins, 1999.
3. Bergonse FN, Cymbalista NC, Nico MM et al: Giant nevus lipomatosus cutaneus superficials: case report and review of the literature. J Dermatol 27: 16-19, 2000.
4. Dotz W, Prioleau P: Nevus lipomatosus cutaneus superficialis. A light and electron microscopic study. Arch Dermatol 120: 376-379, 1984.
5. Köse O, Balo lu H: Nevüs Lipomatosis Süperfisiyalis. T Klin Dermatoloji 7: 208-10, 1997.
6. Birinci Ç, Akbay G, Kapusuzo lu N , Kaur AC, Karabay Y: Nevus Lipomatozus Süperfisyalis. Lepra Mecm 57-61, 1996.
7. Akman A, Yücel A, Gümürdülü D et al: Nevüs lipomatozuslu bir olgu. XIV. Prof.Dr. A. Lütfü TAT ssimpozyumu. Özet Kitab . Ankara, 1999, pp: 106.

8. Park H, Park C, Yi J, Kim T, Kim C: Nevus lipomatosus superficialis on the face. Int J Dermatol 36: 435-7, 1997.
9. Aksoy D, Vahapo lu H, Dereboyu G, Eskio lu F: Nevus Lipomatosus Cutaneus Superficialis (Hoffmann-Zurhelle). Dermatopatoloji Dergisi 4: 112-114, 1995.
10. Mu, Xiao C. Tran, Tien-Anh N: Dupree, Marsha; Carlson, J. Andrew. Acquired vulvar lymphangioma mimicking genital warts. A case report and review of the literature. J Cutan Pathol 26: 150-154, 1999.

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