Nevus Lipomatosus Cutaneus Superficialis (Hoffmann-Zurhelle)

*Presentation of a Case and Review of the Literature*

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The following case report is presented because of the comparative rarity of this nevus. To our knowledge, only eight reports of this condition have appeared in the world's literature, only two in America.

Nevus lipomatoses cutaneus superficialis was the title given by Hoffmann and Zurhelle to a nevoid anomaly consisting of grouped papules and nodules which on histologic examination showed the presence of ectopic fatty tissue in the corium. The second case was described by Robinson and Ellis in 1937. In more recent years cases have been presented by Nikolowski, Kuta, Thöne, Holtz, Blumenthal, and Hering.

Enough similarity exists in the clinical and histologic findings in all reported cases that there can be little doubt that this is an entity. Usually, at least part of the nevus had been present at birth, though there has often been extension of the lesion during childhood. In Holtz's patient, however, the nevus appeared at 18 years of age. Notable is the predilection of this nevus for the region of the pelvic girdle. The reported sites include the buttocks, sacral and coccygeal region, and various surfaces of the upper thighs. The greatest deviation was in the case of Kuta, where the lesion was on the right side near the angle of the ribs.

Typically, plaques measuring up to 8×15 cm. are made up of groups of non-tender papules and nodules, often arranged in linear or systematized distribution, sometimes following the lines of body folds. The papules have been as small as pinhead size, elevated only a millimeter, up to confluent nodules 2×0.5 cm., elevated 1 cm. above the surrounding skin surface. The color is the same as nearby normal skin or pale yellow or yellow-red. The surface is usually smooth but may be irregularly folded or even warty. The consistency is invariably soft, and some of the nodules can be pressed down into the skin in a manner reminiscent of macular atrophy.

Histopathologically important is the presence of fat cells in the middle and deeper portions of the corium. The strands of fat cells are of variable size and may in the aggregate occupy a large part of the corium, being separated only by remnants of collagen in the form of septums. They do not extend into the papillary portion of the corium and may or may not be continuous with the subcutaneous fat. There is often a grouping of these fat lobules around appendages and, particularly, blood vessels.

Epidermal changes, while usually present, appear to be secondary. They consist of hyperkeratosis with follicular plugging, moderate acanthosis with squaring of the rete pegs, and hyperpigmentation of the basal layer, sometimes associated with increased deposition of melanin in the upper part of the corium. The epidermal changes, therefore, resemble those of epidermal nevi.

Considerable discussion has centered around the histogenesis of this nevus. Hoffmann and Zurhelle found the collagen fibers surrounding the masses of fat cells to be degenerated, consisting of fine fibers which stained only weakly with the usual connective tissue stains. The areas of loose con-

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nective tissue stained metachromatically with polychrome methylene blue and crystal violet. In this region the elastic fibers were also fragmented. These authors were led to the conclusion that the deposition of fat is secondary to some primary degenerative change of the connective tissue. This opinion was concurred in by Nikolowski, who found similar degenerative changes and metachromasia in the connective tissue in his cases.

On the other hand, Robinson and Ellis expressed the opinion that the fat cells in the cutis represented a nevoid anomaly and were not the result of degeneration of the collagen. Kúta and Thöne also found little evidence of degeneration of collagen and no metachromasia. The elastic fibers, though fine and fragmented, were present. These authors also interpreted the histogenesis to represent heterotopic development of fat in the corium, with only secondary collagen change.

The most thorough study of this question was carried out by Holtz, who demonstrated, in addition to large mature fat cells, less mature forms comparable to those found in embryonic panniculus adiposus. These apparently arose from the wall of the venous capillary around which the abnormality centered and grew into the surrounding connective tissue, which persisted as very fine collagen, elastic, and reticulin fibers reaching from the unaffected connective tissue to the wall of the central vessel. On staining with crystal violet and thionine, metachromatic substance was found to be adherent to the fibrils. Holtz, therefore, concluded that the primary change is a growing out of primitive “preadipose” tissue from blood vessel walls. Edema and invasion of the surrounding connective tissue result in its destruction. The young adipose cells then progressively take on fat and at last come to be identical with mature cells of the subcutaneous layer. This process corresponds to that described by Wassermann in the embryonic development of normal fatty tissue and in lipomas.

Report of a Case

A woman,* 18 years of age, first noted plaque-like thickening of the skin of the upper portion of the left buttock at the age of 13. This gradually enlarged without symptoms, except possibly very slight itching. There was no treatment. The patient’s general health was good, and there was no evidence of other nevoid disorders.

On the upper portion of the left buttock was a group of pale yellow, slightly elevated and thickened rounded papules 2 to 4 mm. in diameter in an area about 5 cm. in diameter. (Fig. 1)

Histopathologic Findings.—The epidermis shows marked hyperkeratosis of a basketweave variety, and the follicular orifices are dilated and plugged by loose keratin (Fig. 2). The rete is either thinned or acanthotic, with some sharpening of the corners of the rete pegs. The basal layer contains moderate amounts of melanin. The over-all epidermal structure is, therefore, reminiscent of epidermal nevi.

* This patient was presented before the Minnesota Dermatological Society on Feb. 22, 1958.
The papillary corium is essentially normal. Immediately beneath it, extending transversely, is an extensive deposit of fatty tissue which almost completely separates the papillary portion from the deeper parts of the corium. This band is penetrated here and there by strands of collagen. Numerous dilated blood vessels can be seen coursing through the area of fatty tissue, and in some places the fat cells follow the vessels as they ascend toward the papillae, while in the deeper part of the corium fatty tissue surrounds the larger blood vessels as they rise from the hypoderm. The fat-containing cells appear mature. Groups of these fat cells are separated by narrow strands of collagenous connective tissue, the fibers of which are of approximately the same diameter as the collagen fibers in the papillary portion of the corium. They stain slightly less intensely with eosin than the thicker collagen fibers elsewhere in the cuts. Sudan III shows the fat cells in the corium to take the same bright orange stain as the cells of the subcutaneous layer (Fig. 3).

With the Van Gieson stain the collagen fibers appear normal except where interrupted by the fat deposits (Fig. 4). Collagen directly adjacent to the fat cells has normal staining properties, as do the fibers running in the septums between the cells. The Weigert elastic tissue stain shows the elastic fibers to be preserved and, in fact, to run between the individual fat-containing cells in some places. There is no evidence of fragmentation or loss of elastic tissue fibers (Fig. 5).

With the periodic acid-Schiff reaction, no abnormality of the connective tissue or ground substance is noted (Fig. 6). The cell membranes of the fat-containing cells are Schiff-positive; the blood vessels which run through the center of the fat globules are prominent because of the positivity of their endothelial lining.

No metachromasia is noted after staining with toluidine blue.

In summary, the findings in our case support the view of Holtz that the primary disturbance in this nevus is the growth of adipose tissue from the walls of blood vessels and that degenerative changes in the connective tissue, if present, are secondary phenomena. No significant degenerative changes in the connective tissue are demonstrable in our sections.
Comment

This nevus, though undoubtedly rare, has probably escaped recognition in the past. Hoffmann and Zurhelle and Nikolowski have listed previously reported cases which may have belonged to this group. All of these cases were either clinically atypical or not confirmed histologically. This condition is probably most easily confused clinically with other types of connective tissue nevi, sebaceous nevi, or grouped lesions of macular atrophy and histologically with lipoma and perhaps also epidermal nevi. The clinical characteristics of this nevus prevent confusion with lipoma, while standard reference works on dermatologic histopathology are unanimous in stating that lipomas are subcutaneous tumors. Their extension into the skin has not been described to our knowledge.

The predominant localization of these nevi around or near the pelvic girdle has provoked comment by several authors. The reason for it remains unknown. Nikolowski speculated about some peculiarity of the circulation in that region and also pointed out that the gluteal area is subjected to lasting pressure and tension during intrauterine life. Holtz pointed out that this area of the body is “lipophilic,” that is, the fat pad is normally thick here, and this is frequently a site of predilection for other disorders of fat, such as lipodystrophies and dystrophia adiposogenitalis. On the other hand, as Günther pointed out, lipomas are relatively rare in this region.

Summary and Conclusions

An instance of nevus lipomatous cutaneus superficialis (Hoffmann-Zurhelle) occurring in the gluteal skin of an 18-year-old woman is described. This is the third case to be reported in America and the ninth in the world’s literature.

The histopathologic findings in this case support the view that the primary disturbance is an ectopic growth of adipose tissue arising from the blood vessels of the corium, not resulting from fatty degeneration of the connective tissue.

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REFERENCES