

Nevus lipomatosis superficialis of Hoffmann and Zurhelle

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Sir,

Nevus lipomatosis superficialis was first described by Hoffmann and Zurhelle in 1921 [1]. This nevus has been described to be a rare benign hamartoma [2].

Our patient a 20-year-old male who came to us with a multiple raised lesion over the left flank. The lesions were asymptomatic. The lesions were of varying sizes ranging from 1 x 1 cm to 6 x 4 cm. Some of the individual lesions have coalesced to form a single larger lesion. The overlying skin has an irregular surface with some areas showing small openings. He reports that on squeezing these lesions, white cheesy material extrudes out from these openings. The patient has had these lesions since birth however they have been progressively increasing in size in proportion to his growth. No similar history in the family. On examination we observed multiple skin colored to hyperpigmented papules, plaques and nodules present unilaterally on the left flank distributed linearly. The skin over few lesions also had yellowish appearance. The lesions were well defined with varying sizes and shapes (1 x 1cm to 6 x 4 cm) with an irregular surface and margin. (Fig. 1) Few of the papules and plaques have coalesced to form a larger lesion. Some areas show a cerebriform surface and comedo like openings. Most of the lesions were sessile with few being pedunculated. The lesions were non tender. White cheesy material could be expressed out from the comedo like opening on applying pressure. Dermoscopic evaluation was done using a DermLite DL4. Dermoscopy showed areas of cerebriform pattern with sulci and gyri, yellow structureless areas, comedo like opening and pigment network (Figs. 2 and 3). A 3.5 mm punch biopsy was taken from the lesion and sent for histopathological examination. The biopsy

report showed, varying proportion of mature adipose tissue around sub papillary vessels. Dermal collagen appears thickened and vascularity appear greater than normal. Boundary between dermis and hypodermis was ill defined (Fig. 4). Hence a diagnosis of Nevus lipomatosis superficialis of Hoffman and Zurhelle was made.

The patient was assured the benign nature of the lesion and was explained the option of surgical removal. He however was not willing for any surgical procedures at the time.

The word nevus is derived from Latin meaning “maternal impression”. It is used synonymously with hamartoma which is derived from Greek word hamartia meaning “error”. The nevus lipomatosis superficialis is a fat nevus. It has two known forms the classical type and the solitary type. The classical type was described by Hoffman and Zurhelle [1], and hence it has been named after them. The lesions are multiple, soft, nontender, pedunculated, cerebriform, yellowish or skin-colored papules or nodules [3]. The lesions may coalesce. The most common site is the pelvic girdle area, i.e., the involvement of one hip or buttock [3-5]. The trunk and abdomen are rare sites of manifestation⁵ as was in our patient. The onset of the lesion is usually at birth or at infancy. The solitary lesions have a later onset with a predilection to the trunk [5]. The nevi maybe associated with comedo-like lesions and hypertrichosis over the nevus, angiokeratoma of fordyce, cafe-au-lait or vitiligo-like macules, hemangioma, and basal cell carcinoma [6]. Our patient had comedo-like lesions. It is considered to be a nevoid anomaly with the ectopic fat cells derived from the perivascular mesenchymal tissue. Histopathologically it has groups and strands

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Figure 1: Skin colored to yellowish papules, nodules and plaques over left flank with comedo like openings.



Figure 3: Dermoscopy Green arrow- Yellow structureless area. Image also shows comedo like openings and cerebriform surface.

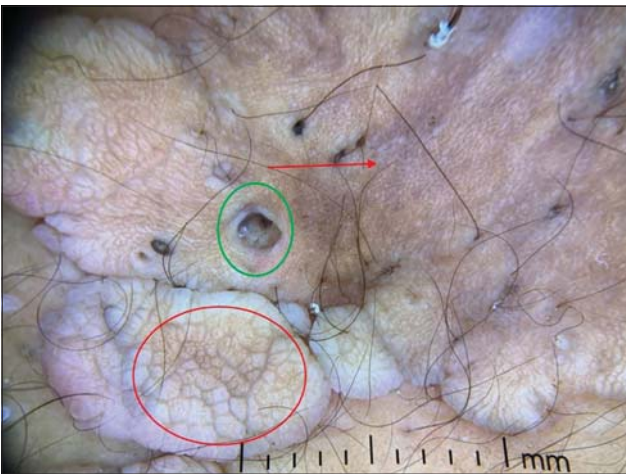


Figure 2: Dermoscopy- Red circle- cerebriform pattern of sulci and gyri; Green circle – comedo like opening; Red arrow- Pigment network.

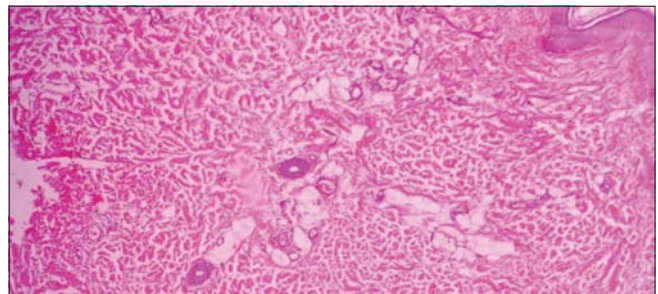


Figure 4: Histopathology image showing mature adipocytes in the dermis in the perivascular location.

of mature fat cells which are found embedded among the collagen bundles of the dermis, often as high as the papillary dermis. When there is large amount of the mature fat cells distributed throughout the dermis the hypodermis and dermis cannot be delineated [5].

Dermoscopy of nevus lipomatosis superficialis is not known to have any distinctive features. Most commonly seen findings are the yellow structureless areas due to the fat deposition and the cerebriform surface due to its irregular surface and comedo like openings. These findings may be present in other conditions like seborrheic keratosis [7].

Differential diagnosis to be considered include, nevus sebaceous, nevus comedonicus, focal dermal hypoplasia, connective tissue nevus, neurofibroma, and acrochordons.

The condition is benign hence surgical removal is for cosmetic purposes.

CONCLUSION

The above case has been reported due to the unusual characteristics observed in this benign hamartoma. Our patient had a presentation in the trunk. The lesion also showed comedo like openings on its surface. Diagnosing this condition is important as it is completely benign and the patient can be assured of the same.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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