Case Report

Nevus lipomatosus cutaneous superficialis (Hoffman-Zurhelle) over lower back: a rare presentation

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ABSTRACT

Nevus lipomatosus cutaneous superficialis is a rare benign local malformation of the skin characterized by ectopic adipocytes in the dermis. It presents as either a classical type known as Hoffman-Zurhelle or the solitary type. We report a case of the classical type with multiple soft, non-tender, cerebriform, skin-colored papules and nodules over the lower back in a 16 years old female.

Keywords: Adipocytes, Ectopic adipose tissue, Nevus lipomatosus cutaneous superficialis

INTRODUCTION

Nevus lipomatosus superficialis is a rare developmental disorder characterized by isolated ectopic deposition of adipose tissue in the dermis favoring the site of the pelvic girdle. Hoffman and Zurhelle first reported the case in the year 1921.¹ We report a case of classical Hoffman-Zurhelle type of nevus lipomatosus in a 16 years old girl.

CASE REPORT

A 16 years old girl presented with multiple skin-colored nodules on her lower back since birth. The lesions started as multiple small, skin-colored papules and nodules with a smooth surface, which gradually increased in size to attain present size (Figure 1 a and b). The lesion was asymptomatic, with only concern of unsightly appearance. No family history of similar lesions was noted. Physical examination revealed multiple, soft, skin-colored papules and nodules of sizes ranging from 0.3×0.3 cm to 2.0×1.5 cm on the lower back. Histopathological examination revealed epidermis with hyperkeratosis. Dermis shows mature adipose tissue. These findings are suggestive of the diagnosis of nevus lipomatosus.

DISCUSSION

Nevus lipomatosus superficialis is a rare skin malformation characterized by the presence of ectopic mature adipose tissue in the dermis.¹² The proportion of the fatty tissue varies from 10-50% of the dermis.³ Clinically, it is classified into the classical Hoffmann-
Zurhelle form and the solitary form. The classical type, first reported by Hoffman and Zurhelle, consists of multiple, soft, non-tender, pedunculated, cerebriform, yellowish, or skin-colored papules or nodules usually situated on the pelvic girdle area in a zonal pattern and can occur at birth or during first three decades of life. Rare involvement of the face or scalp has been reported. They are almost invariably asymptomatic, although occasionally, ulceration may occur. The solitary form of NLCS usually appears during the third to sixth decades of life as a single papule or nodule without a favored location. There is no evidence of familial tendency or sex predilection in either of the clinical types. There are reports of coexisting café-au-lait macules, leukodermic spots, overlying hypertrichosis, and comedo-like alteration. The exact pathogenesis of NLCS is not known. Few proposed hypotheses are: dermal adipocytes may originate from pericytes, NLCS may be a connective tissue nevus, and deletion of 2p24 in NLCS thus supporting role of genetic factors in the development of NCLS.

Clinically, the differential diagnosis includes neurofibromatosis, lymphangioma, nevus sebaceous, hemangioma, skin tag, or fibroepithelial polyp, all of which can be distinguished histologically. Microscopically, islands of mature adipose tissue are present in the dermis interposed among the collagen bundles and comprising more than 50% of the dermis. Treatment, though not necessary, is done for cosmetic reasons, and simple surgical excision is the best choice. Systemic abnormalities and malignant alterations have not been associated with this abnormality. No significant complications are observed in the majority of cases. Rarely, foul-smelling discharge can occur due to ulceration, and in very rare cases, the tumor may recur after surgical excision.

CONCLUSION

Nevus lipomatosus cutaneous superficialis is a rare benign developmental abnormality with the presence of ectopic mature adipose tissue in the dermis. Early recognition enables for more conservative resection of the tumor for cosmetic purposes.

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REFERENCES
