



NAEVUS LIPOMATOSIS SUPERFICIALIS - A RARE CASE REPORT LAKSHNA S

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Abstract : Naevus lipomatosis superficialis (NLCS) is a rare type of connective tissue naevus. It is a rare idiopathic benign hamartoma. It is characterized by presence of ectopic mature adipose tissue within the collagen bundles of dermis. It clinically manifests as solitary or multiple papules or plaques on skin. Here we report a case of 11 year old female child with multiple soft non tender cerebriform skin coloured papules and plaques in the left hip region. We report this case as it is an uncommon benign lesion to highlight its clinical picture, histopathological features and differential diagnosis.

Keyword : naevus lipomatosis superficialis , hamartoma , cerebriform.

INTRODUCTION

Nevus lipomatosis cutaneous superficialis (NLCS) is a rare benign hamartomatous disorder characterised by isolated mature adipose tissue in the dermis¹. It has predilection to the pelvic girdle. In 1921, Hoffmann and Zurhelle described the first case of NLCS in a 25 year old man who presented with multiple soft nodules on the gluteal region². A case of nevus lipomatosis superficialis is reported.

CASE REPORT

A 11 year old female child presented with complaints of asymptomatic skin lesions on left hip region since 8 years of age. No family history of similar lesions. Physical examination revealed multiple non tender skin coloured papules and plaques with wrinkled cerebriform appearance (FIGURE 1). The excision biopsy was submitted for histopathology.



FIGURE 1-Clinical photograph shows multiple cerebriform skin coloured nodules and plaques.

Gross appearance - A specimen of skin with subcutaneous tissue measuring 10X8X4 cm. External surface was nodular with cerebriform wrinkled skin appearance.(FIGURE 2). On cut section the extension of fat was seen close to epidermis and junction between dermis and subcutaneous tissue was blurred (FIGURE 3).



FIGURE2: Gross-External surface shows skin with subcutaneous tissue with cerebriform wrinkled skin appearance



FIGURE 3:Gross- Cut surface shows extension of fat close to epidermis and blurring of junction between dermis and subcutaneous tissue

Histopathological examination revealed an acanthotic epidermis with flattened rete ridges (FIGURE 4). Both the papillary and reticular dermis contained scattered lobules of adipocytes entrapped between bundles of dermal collagen fibers (FIGURE 5). The lobules of fat were mainly localized around blood vessels (FIGURE 6). Increased vascularity was observed in the ectopic dermal adipose tissue. The ectopic adipose tissue had no connection with the underlying subcutaneous fat. The pilosebaceous follicles in the dermis were reduced. Based on the clinical and histopathological features, a diagnosis of Naevus lipomatosis cutaneous superficialis was made.

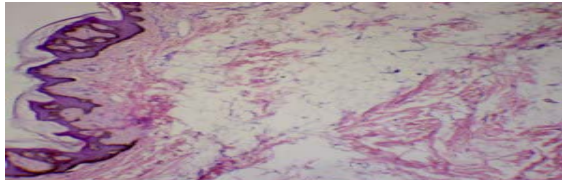


FIGURE 4:Low power view shows acanthotic epidermis with mature adipose tissue seen in the dermis (Haematoxylin & Eosin stain)

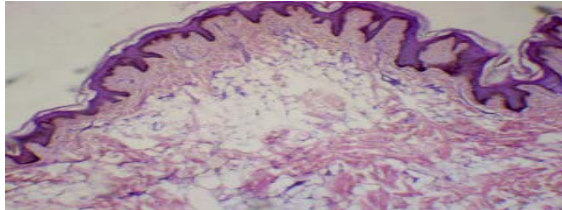


FIGURE 5:Low power view shows mature adipose tissue in the dermis with entrapped collagen bundles. (Haematoxylin & Eosin stain)

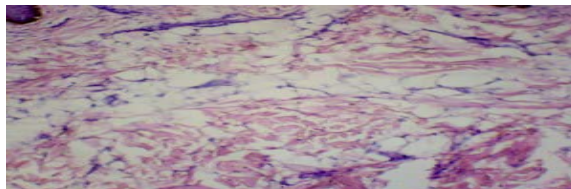


FIGURE6:High power view shows aggregates of mature adipocytes around subpapillary blood vessels. (Haematoxylin & Eosin stain)

DISCUSSION

Naevus lipomatosus cutaneus superficialis (NLCS) is classified into two subtypes (classic or multiple form and solitary) based on clinical presentation. The Hoffmann-Zurhelle form, or the classic form, characterized by multiple soft, non-tender papules and nodules which usually coalesce to form plaques. The classic form has a predilection for the gluteal, pelvic and lower back regions. The lesions are present at birth or develop in the first two decades of life. The solitary form can present as a single pedunculated or dome-shaped papule or nodule or rarely a plaque¹. Pedunculated lipofibroma is seen in fifth decade³. There has been no evidence of correlation with sex or ethnicity⁴. As the solitary form shows clinical and pathological features that differ from the classical type, it is also referred to as pedunculated lipofibroma⁵. The solitary form is predominantly found on the buttocks and thighs⁶, although it may occur at unusual sites like scalp, axilla, knee, ear, eye and palm^{3,7,8}. The main histopathological abnormality in either type of NLCS is ectopic adipose tissue in the upper dermis and not connected with the fat of underlying subcutis. The proportion of the dermal fat is variable, ranges from less than 10% of the dermis to over 50%^{1,4}. When the lesion is small, the fat is usually localized around the subpapillary blood vessels^{1,6}. Excessive, loose, or irregular organization of the connective tissue has been noted in many cases. In our case, there was thickening of dermal collagen and the fat was mainly seen around dermal blood vessels. The epidermal changes are variable. There is often some undulation with acanthosis and even mild papillomatosis and there may be mild hyperpigmentation of the basal layer. The changes may resemble those of an epidermal nevus¹. There are also

abnormalities in the other connective tissue components of the dermis which includes thickening of the collagen bundles and increase in deeper elastic tissue. There is an increase in the number of fibroblasts and blood vessels in the papillary dermis, and also of mononuclear cells, including mast cells in the dermis¹. Blood vessels are also increased in the ectopic dermal adipose tissue. Pilosebaceous follicles are often reduced¹. However in our case, increased vascularity was observed only in the ectopic adipose tissue. The exact pathogenesis of NLCS is unknown. It was thought to be due to deposition of fat secondary to degenerative changes in connective tissue. This theory is supported by Hoffmann, Zurhelle and Nikolowski⁹. However subsequent study failed to support this theory. In 1955, Holtz thought preadipose tissue was derived from dermal blood vessels. In 1980, the electron microscopic finding by Raymond et al strongly supported Holtz's theory. They confirmed the perivascular origin of young adipocytes and differentiation into mature fat^{10,11}. Naevus lipomatosus cutaneus superficialis (NLCS) should be differentiated from nevus sebaceous, focal epidermal hypoplasia, dermal variant of spindle-cell lipoma, benign papillomas like acrochordons and fibroepithelioma of Pinkus¹. Histopathological examination usually helps in the differentiation. NLCS contains mature adipocytes, but no skin appendages in the dermis. Nevus sebaceous and other benign papillomas contain skin appendages, but no adipocytes in the dermis. Solitary form of NLCS has a broad base when compared to fibroepithelioma of Pinkus. Dermal collections of adipocytes on histopathological examination are also present in old nevocellular nevi and some melanocytic nevi. However, the presence of nevus cells sometimes occupying a small area of the lesion helps in the differentiation. Focal epidermal hypoplasia (Goltz syndrome) also has fat in the dermis, but in this condition there is extreme attenuation of the collagen and associated with skeletal abnormalities^{1,12}. The dermal variant of spindle-cell lipoma contains more spindle shaped cells as well as a fibromucinous stroma¹. Treatment for NLCS is not necessary other than for cosmetic reasons⁴. Systemic abnormalities and malignant changes have not been associated with NLCS. Excision is curative and recurrence after surgery is rare⁴. In our case, the lesion was completely excised for cosmetic purpose.

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