Nevus Lipomatosus Cutaneous Superficialis

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ABSTRACT

Nevus lipomatosus cutaneous superficialis (NLCS) is an uncommon benign hamartomatous skin lesion defined by the presence of aggregates of mature adipose tissue among the collagen bundles of dermis. It was first reported by Hoffman and Zurhelle in 1921. It is classified into two types. The classic form with multiple soft,pedunculated,cerebriform papules and nodules that coalesce into plaques and the solitary form that consists of solitary papule or nodule. The classical NLCS is mostly reported to involve pelvic or gluteal region. We report a case of 17 years old girl with this classic form of the rare skin malformation.

Keywords: Nevus lipomatosus superficialis, adipocytes, hamartomatous skin lesion.

INTRODUCTION

Nevus lipomatosus superficialis is a rare benign idiopathic skin malformation characterised by ectopic mature adipose tissue in the dermis.The proportion of the fatty tissue varies greatly from greater than 50% to less than 10% of the dermis[1]. Clinically it is classified into two types,the classical Hoffman – Zurhelle or multiple form and the pedunculated solitary form. The classic type first reported by Hoffman and Zurhelle, consists of multiple, soft, non-tender, pedunculated, cerebriform, yellowish or skin coloured papules or nodules usually situated on the pelvic girdle are in a Zonal pattern and occur at birth or during 1st three decades of life[2]. The solitary form usually occurs after the age of 20 yrs,presents with a single nodular lesion with no particular predilection sites[2,3]. Here we present a case of 17 yr old girl with classical NLCS on the gluteal region.

CASE REPORT

A 17 years old girl presented with a well defined multiple ,smooth, soft and non tender nodular growths over the gluteal region for the preceding 3years.There were no other complaints. Examination revealed multiple skin coloured, cerebriform nodular lesions measuring 3.5 X 2 cm over the gluteal region [Figure 1]. There was no tenderness or ulceration or induration. There was no regional lymphadenopathy. Systemic examination normal. Routine laboratory examination of blood was within normal limits. Histopathological examination of the incisional biopsy specimen showed epidermis with aggregates of mature adipocytes in the papillary dermis and around eccrine glands and between the collagen bundles [Figure 2&3]. Based on the clinical and histopathological features ,a diagnosis of the classic variety of NLCS was made.

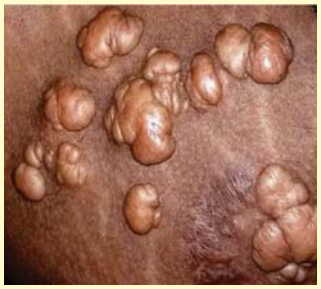

Figure 1: Multiple Cerebriform Nodules Over Gluteal Region

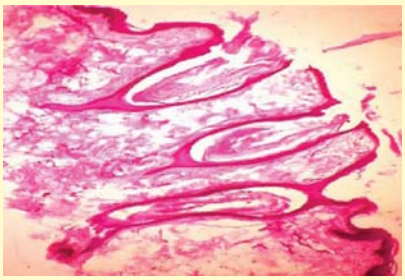

Figure 2 : H&E section (10 X) showing adipocytes in papillary Dermis

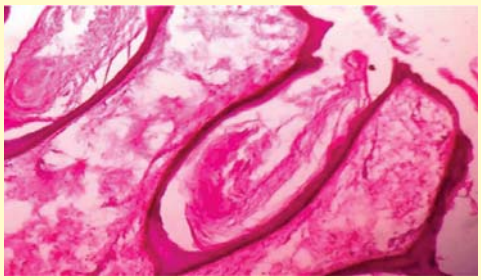

Figure 3 : H&E Section showwing (40X) adipocytes extending upto epidermis.

DISCUSSION

Nevus lipomatosus superficialis is an uncommon benign hamartomatous skin lesion. The classic type of NLCS is usually unilateral as seen in our case. The classic variety of NLCS is either present at birth or can arise at any other time within in first two decades of life[4,5,6]. There are sporadic case reports of coexistent anomalies in the form of café-au-lait macules and scattered leucoderma. Increased hairness and comedo-like lesions have also been reported[7]. The histopathology of NLCS usually shows a normal or slightly attenuated epidermis associated with dermal proliferation of mature adipocytes in the reticular dermis that may extend to papillary dermis . The adipocytes most commonly form small aggregates around blood vessels or eccrine glands, but may also be present as solitary adipocytes between collagen bundles. The proportion of dermal fat is variable, ranging from less than 10% to more than 50%[4]. The adipocytes may show conection to the underlying subcutaneous fat or be separated from the subcutis by collagen. Less commonly,spindle cells representing immature fat cells may also be present. NLCS should be differentiated from nevus sebaceous, skintags, neurofibroma, lymphangioma and focal dermal hypoplasia (Goltz syndrome). Histopathological examination usually helps in the differentiation. No fat cells are present in the dermis in the case of skin tags. Simillar dermal collections of adipocytes on histopathological examination are also present in some melanocytic nevi, pedunculated lipofibromas and in Goltz syndrome. Lipofibromas contain fat cells, but no skin appendages in the dermis. In the case of Goltz syndrome, there is absence of collagen in the atrophic dermis and skin appendages are absent. The precise etiopathogenesis of NLCS is not known. There is no specific explanation behind the predilection of the lesions for the pelvic area[7,8,9]. Deposition of adipose tissue may be caused by the degenerative changes in dermal collagen and elastic tissue[10,11,12].

The proposed pathogenesis of NLCS includes adipose metaplasia dermal connective tissue, developmental displacement of adipose tissue, alternatively, the lesions could be explained by possible origin of adipocytes from pericytes of dermal vessels[10].

Treatment is not necessary other than for cosmetic reasons. Systemic abnormalities and malignant changes have not been associated with NLCS. Excision is curative and recurrence after surgery is rare.

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