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CASE REPORT

NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS – CLASSICAL FORM – A CASE REPORT

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ABSTRACT

We report Nevus Lipomatosus Cutaneous Superficialis (NLCS) which is considered a rare entity. Two types are recognized, the classical form with multiple, soft, pedunculated, cerebriform papules and nodules that coalesce into plaque and the solitary form consisting of solitary papule or nodule. Histologically it is characterized by the presence of mature ectopic adipocytes in the dermis

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INTRODUCTION

Nevus Lipomatosus Cutaneous Superficialis is an uncommon benign hamartomatous condition characterized by the presence of mature ectopic adipocytes in the dermis. It was first reported by Hoffman and Zurhelle E. in (1921). Clinically it is classified into two forms (Yap, 2009). Classical form is characterized by groups of multiple non tender, soft pedunculated cerebriform, yellowish color papules, nodules or plaques. The other form of NLCS classically manifested as a solitary, sessile papule (Yap, 2009). Classical form most commonly seen at gluteal and pelvic regions. We here with report two cases of classical form of NLCS.

Case Report

- A 55 year male presented with swelling in gluteal region since 1 year and gradually increased in size. Grossly single pedunculated globular mass.
- A 40 year female presented with over left buttock since one and half year.

Grossly single papillary mass with external cerebriform appearance.

In both cases no similar lesions were noted elsewhere, there was no family history of similar lesions. History of trauma was not present.

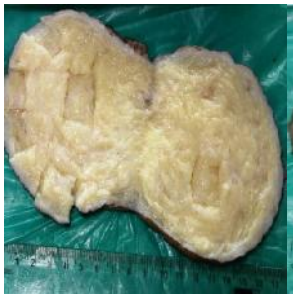

Fig. 1.

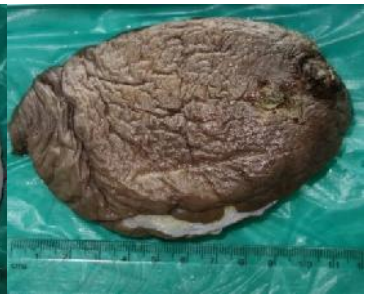

Fig. 2.

Dome shaped mass. External Surface – Cerebriform appearance

Cut Surface – Yellowish white and greasy

Histopathological examination of both specimen shows lobules of mature ectopic adipocytes with intervening dense collagen bundles in the dermis and at places surrounds perivascular and periadnexal area. There is no connection of these ectopic adipocytes with subcutaneous fat.

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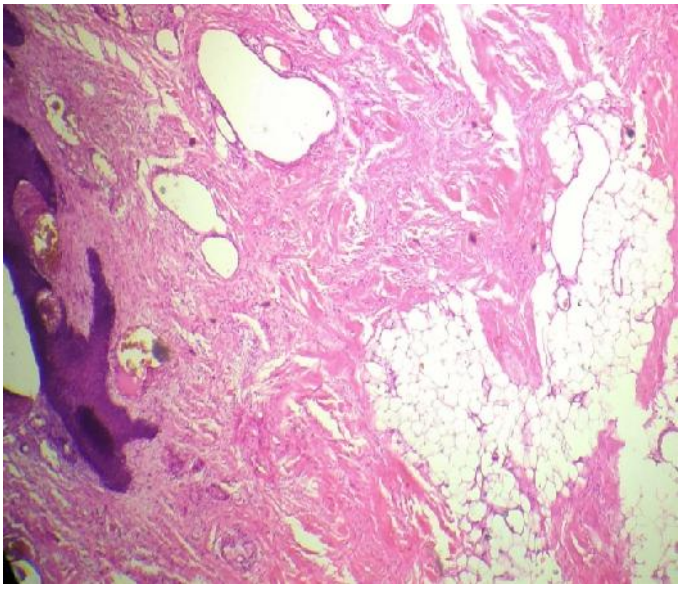

Fig. 3 & 4. Histopathology showing normal epidermis with islands of mature adipocytes in dermis

DISCUSSION

NLCS is a commonly diagnosed in pediatric cases but can appear later in life, Sex predilection or familial trend are not observed in this disorder (Hatori, 2003). In the classical type, lesions are congenital as they develop usually during first two or three decades of life (Yap, 2009). It consist of multiple papules, yellowish in color and having cerebriform external appearance. The lesions are slow growing and can reach a large size if left untreated. The most common sites are the pelvic girdle, the lower trunk, gluteal region and the thigh. The second clinical pattern is a solitary papule or nodule, occurring at any site and also called pedunculated lipofibroma (Lipofibroma, 1994). NLCS have an asymptomatic course. Occasionally NLCS may ulcerate after external trauma or ischaemia. NLCS has been reported in association with other cutaneous disorders; follicular papules and hypertrophic pilo-sebaceous units (Inoue, 2002), Angiokeratoma of Fordyce (Al-Mutairi, 2006), scattered leukoderma, café-au-lait macules (Ghosh, 2010), and hemangioma (Al-Mutairi, 2006). In our study, no associated skin abnormalities were present. NLCS should be clinically differenciated from nevus sebaceous, neurofibroma, lymphangioma, focal dermal hypoplasia, cylindroma, trichoepithelioma, and angiolipoma. Histopathological evaluation is required for diagnosis showing ectopic mature adipocytes, intermingled with collagen bundles, and proliferating around the periadnexial adventitial dermis and the perivascular area. Mature fat cells proliferate in reticular dermis and may extend to the papillary dermis (Ghosh, 2010; Ioannidou, 2001; Kaw, 2005).

The pathogenesis of NLCS remains unknown. Several theories have been proposed: Hoffman and Zuhrelle postulated that fat deposition in the dermis is secondary to degenerative changes (metaplasia) in the connective tissue (Buch Archana, 2005). Treatment is usually not necessary unless for cosmetic reasons. The treatment of choice is surgical excision, which is curative, and post surgical recurrence is rare.

Conclusion

Early recognition enables more conservative resection of the tumor and less invasive reconstruction of the defect. NLCS is a rare skin malformation. The physician should aware of this rare tumor which can become extremely large if untreated.

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