NEVUS LIPOMATOSUS CUTANEOUS SUPERFICIALIS - A RARE CASE REPORT

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ABSTRACT
Benign lipomatous tumours of the mesenchyme represent a common group of neoplasms which pose little diagnostic difficulty. Hamartomatous lesions can also arise from or intimately associated with adipose tissue. Nevus lipomatosus cutaneous superficialis (NLCS) is a rare cutaneous hamartomatous lesion. Usually presents in classical form also called Hofman-Zurhelle nevus, other form being the solitary form. Herein we present this case in a seventeen year old female, a classical form of NLCS who presented with multiple skin coloured, papular lesions over the buttocks from 3 years, with recent increase in size of the lesions.

Key Words: Adipocytes, Cutaneous Neoplasms, Cerebriform, Gluteal, Lipomatosus and Papules

INTRODUCTION
NLCS is a hamartomatous lesion with presence of mature and ectopic adipocytes in the upper dermis. It was first reported by Hoffman and Zurhelle in 1921 and is regarded as connective tissue nevus by most authorities (Ragsdale, 2009). There are two forms: classical and solitary. Classical form involves pelvic, gluteal region or upper thigh. Clinically, they present as group of multiple skin coloured or yellowish papules, nodules, comedo like lesions or Cerebriform lesions. No gender predilection noted. Rare sites are shoulder, scalp, abdomen, back and thorax (Dhamija et al., 2012). The classical form is usually unilateral with band like linear or zosteriform distribution. The lesion continues to progress for many years. It may ulcerate with foul smelling discharge. Coexisting anomalies are café au-lait patches and leukoderma. The solitary form usually occurs after the age of twenty years, presents with a single nodular lesion with no particular predilection site. There is no association with systemic abnormality or malignant alterations (Goucha et al., 2011). Differential diagnosis includes Nevus sebaceous, Fibrolipoma, skin tag, Neurofibroma, Lymphangioma, Golts syndrome and Melanocytic nevi. Etiopathogenesis is not understood but deposits occur due to degenerative changes in dermal collagen and elastic tissue i.e theory of Adipose metaplasia has been thought of treatment consists of surgical excision for cosmetic reasons. No recurrences noted (Pujani et al., 2014).

CASE REPORT
A 17 year old female presented with multiple skin coloured, papular lesions over both buttocks since 3 years. Started as few millimeters sized lesions and progressed to the present state. No history of itching, pain or any discharge. Local examination revealed papules, lobulated and plaque lesions of size ranging from 2mm to 5cm which were non-tender. Multiple comedowns noted over surface of few plaques. Systemic examination revealed no abnormality. All Laboratory investigations were within normal limits. The suggested clinical diagnoses were Dermolipoma and Xanthoma. Patient underwent surgical excision.

Figure 1: Multiple skin coloured papules and nodules over the gluteal region
**Gross:** we received multiple skin covered soft tissue bits measuring 2x1 cm size. Cut surface grey brown to grey white. Microscopy showed normal to focally attenuated epidermis. Dermis showed lobules of mature adipocytes in the reticular dermis extending into papillary dermis. They also formed small aggregates around blood vessels and adnexae. Hair follicles were few. Immature fat cells also seen at focal areas.

![Figure 2](image2.png)

**Figure 2:** Section showing epidermis and dermis with proliferation of mature adipocytes in the reticular dermis extending into papillary dermis (H&E 4x)

![Figure 3 & 4](image3.png)

**Figures 3 & 4:** Sections showing mature adipocyte clusters encircling the adnexal structures (H&E 10x, 40x)

**DISCUSSION**

NLCS is a nevoid accumulation of mature adipocytes and is considered to be a connective tissue nevus. Nevi of connective tissue are divided into adventitial and reticular nevi. Nevi of reticular connective tissue are situated on the trunk and consist of faulty distribution of fat. NLCS can be of 2 types: classical and solitary forms. A rare variant has been described called Michelin tire baby syndrome with autosomal dominant inheritance (Burgdorf et al., 1982). The classical form also known as Hoffman Zurhelle nevus consists of clustered, pedunculated, cerebriform skin coloured or yellowish nodules of soft, fleshy consistency usually involving the trunk in a linear pattern. The solitary form is also called pedunculated lipofibroma as coined by Mehregan et al. (2015). Rare cases show ulceration or gluteal hypertrophy. Associated hair follicle abnormalities can be noted like reduction in number and trichofolliculoma formation are also noted (Khandpur et al., 2009).

Several theories have been proposed for its etiopathogenesis including adipose tissue metaplasia during the course of degenerative changes in dermal connective tissue and mesenchymal perivascular cells transforming into Littoral lipoblasts and then into mature adipocytes. 2p24 deletion has also been noted (Wollina, 2013).
CONCLUSION
NLCS can have a variable clinical presentation, classical type being the most common type as in our case. It is usually asymptomatic. Surgical treatment is the cornerstone for these cases. Rare association with other malignancies should be remembered.

REFERENCES
Wollina U (2013). Photoleter to the editor - Nevus lipomatosussuperficialis (Hoffmann-Zurhelle). Three new cases including one with ulceration and one with ipsilateral gluteal hypertrophy. Journal of Dermatological Case Reports, 30 7(2) 71-73.