

NEVUS LIPOMATOSIS CUTANEOUS SUPERFICIALIS – A RARE CASE REPORT IN A TERTIARY CARE CENTRE.

Otorhinolaryngology

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ABSTRACT

Nevus lipomatosis superficialis (NLS) is an uncommon benign hamartoma characterized by ectopic adipose tissue within the dermis, typically presenting as multiple soft, pedunculated papules or nodules, predominantly in the pelvic and gluteal regions. This case report describes a rare instance of NLS localized to the left ear lobe in a 55-year-old female. The patient presented with a painless, slow-growing nodule on the left ear lobe, which was confirmed through histopathology to be NLS. Surgical excision was performed, and the patient had an uneventful recovery with no recurrence observed at a six-month follow-up. This case underscores the importance of considering NLS in the differential diagnosis of soft tissue lesions on the ear lobe and highlights the favourable prognosis following surgical treatment.

KEYWORDS

Benign hamartoma, nevus lipomatosis superficialis, adipose tissue, pedunculated papule.

INTRODUCTION

A Hamartoma is a noncancerous/benign tumor-like mass composed of mix of mature tissue or cells that are present in abnormal proportions or show a disorganized arrangement[1]. Nevus lipomatosis superficialis (NLS) is also known as Dermolipoma, it is an uncommon benign hamartoma characterized by the presence of aggregates of mature ectopic adipose tissue among the collagen bundles of the dermis. They are native to the area where the growth occurs. This neoplasia is usually present at birth or in childhood with no familial tendency nor sex predilection. It is of 2 forms: a) classical type b) solitary type. It typically presents as multiple, soft, and pedunculated papules or nodules. While it is usually observed in the pelvic and gluteal regions, its occurrence on the ear lobe is extremely rare. We present a unique case of NLS localized to the left ear lobe in a 55-year-old female.

Case Report

A 55-year-old female presented with a painless, slow-growing mass on her left ear lobe over the past several years. On physical examination, the lesion was a soft, skin-colored, pedunculated nodule measuring approximately 2 x 2 cm in diameter. The overlying skin appeared normal with no signs of ulceration or inflammation as shown in **fig 1**.



Figure 1: Shows Pedunculated Nodule Of Left Ear Lobe.

The lesion was excised and the specimen is shown as below. A biopsy of the lesion was performed and histopathology (HPE) revealed sheets of melanocytes with minimally pleomorphic round to ovoid vesicular nuclei, conspicuous nucleoli and cytoplasmic melanin pigment, cells spindled out and forming storiform pattern focally. Sheets of mature fatty tissue is admixed with melanocytes in the upper dermis. Cells percolate through the existing sebaceous glands and eccrine glands. Deeper tissue shows lesion admixed with fatty tissue. Mitosis is occasional. No junctional activity noted as shown in **fig 2**.

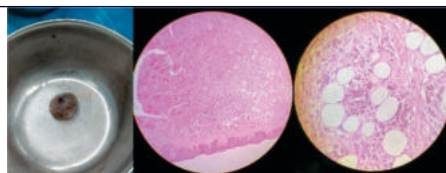


Figure 2: Showing Specimen And Histopathology

Further confirmation was done by ImmunoHisto Chemistry with S100 and HMB45 marker in which S100 showed diffuse strong cytoplasmic and nuclear positivity compatible with melanocytic differentiation and HMB45 showed diffuse moderate cytoplasmic staining compatible with melanocytic differentiation as shown in **fig 3**

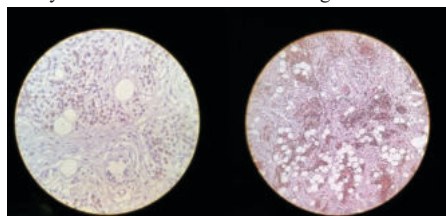


Figure 3: Showing HMB45 and S100 staining positive for melanocytic lesion

There were no atypical features or evidence of malignancy. Given the benign nature of the lesion, surgical excision was recommended. The lesion was completely excised under local anaesthesia and the patient had an uneventful recovery and wound site healthy as shown in **fig 4**. No recurrence was observed at a 6-month follow-up.



Figure 4: Showing Healthy Scar Tissue

DISCUSSION

Nevus lipomatosis superficialis (NLS) is an uncommon cutaneous hamartoma that was first described by Hoffmann and Zurchelle in 1921[2]. NLS is a rare entity, especially when localized to the earlobe. NLS can be subdivided into two forms: the classical (or multiple) type and the solitary type. The classical type is generally seen in early childhood and presents as multiple, soft, yellow or skin coloured papules or often pedunculated nodules with smooth or wrinkled appearance located on the pelvic, gluteal, upper thigh regions and lower back regions[3]. The solitary type, which tends to present above 20years of age, is a rare form typically manifests as a single lesion characterized by dome- shaped sessile papule or nodule and can occur at various sites on the body like face, scalp, eyelids, and clitoris. NLS is a rare condition, with fewer than 100 cases reported in the literature[4]. The overlying skin is often normal, though it may show signs of hyperpigmentation or hyperkeratosis[5]. Furthermore, Café au lait macules, leukodermic spots, overlying hypertrichosis and comedo like alterations can co exist. The pathogenesis remains unclear, though it is hypothesized to result from developmental anomalies, post traumatic changes and degenerative processes[6]. Several theories have been proposed. One such theory is multiple type of NLCS arises from adipose metaplasia in the course of degenerative changes in the dermal connective tissue. Another theory is that the adipocytes would represent a true Nevus from the developmental displacement of adipose tissue. A third proposal is that the mature adipocytes grow from mononuclear cells differentiating into lipoblasts in a perivascular area. The histopathology of NLCS shows clusters of ectopic mature adipose tissue among collagen fibers in the dermis with no connection to the subcutaneous fat tissue. These ectopic adipocytes contain large intracytoplasmic lipid vacuoles and are often associated with vascular structures. In some cases there is an increased an increased density of collagen fibers, fibroblast, and a perivascular infiltration of mononuclear and spindle-shaped cells. The epidermis exhibit acanthosis, basket weave hyperkeratosis, increased basal pigmentation, and obliteration with focal elongation of rete ridges. Adnexal structures may be unaffected or reduced in some cases and may show perifollicular fibrosis .

DIFFERENTIAL DIAGNOSIS
1. Lipoma
2. Keloid
3. Skin tags
4. Neurofibromas
5. Connective tissue Nevus
6. Nevus sebaceous
7. Focal dermal hypoplasia (Goltz syndrome)
8. Lymphangioma
9. Hemangioma
10. Fibroepithelial polyp

In the case of Goltz syndrome, besides the cluster of adipocytes in the dermis, there is an extensive attenuation of collagen in the atrophic dermis and skin appendages are absent[7]. The prognosis is excellent, with a very low risk of recurrence following excision. Treatment is indicated only for cosmetic reasons since malignant degeneration and systemic abnormalities are rare. In cases where the lesion is extensive or involves cosmetically sensitive areas, such as the face or ear, more conservative treatment options may be considered[8]. These may include intralesional corticosteroid injections or laser therapy, though their efficacy is less well established[9]. Due to rarity of recurrent lesions, surgical excision is the treatment of choice, offering both diagnostic confirmation and therapeutic resolution.

CONCLUSION

This case highlights the importance of considering NLS in the differential diagnosis of soft tissue lesions on the ear lobe. Treatment, though not necessary, is done for cosmetic reasons and simple surgical excision appears to be curative as we havedone in this case. Lesions are generally non progressive and systemic abnormalities and malignant alterations have not been associated with this abnormality.Although rare, awareness of this condition can facilitate prompt diagnosis and management, ensuring favourable outcomes for patients.

Abbreviations:

NLS : Nevus lipomatosis superficialis. Fig: Figure

HPE: Histopathology examination.

Conflicts Of Interest/competing Interest: Nil

Funding: Nil.

Ethics Approval: Not applicable

Consent To Participate: Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

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