Nevus lipomatosus superficialis, an unusual case report

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Abstract

Nevus lipomatosus superficialis is a rare hamartomatous lesion of skin, also named as pedunculated lipofibroma. This entity is unknown to many primary care physicians. Hence, many times, it is not diagnosed and misinterpreted as any of its differential diagnoses like lipofibromas, skin tags, hemangioma, lymphangioma, and focal dermal hypoplasia. Histopathologically, it is characterized by the presence of clusters of mature fat cells in the dermis. Here we report a case of an 11-year-old male who presented with a lower back swelling since birth (4.8 x 3.6 cm) which gradually increased over time and became lobulated, pedunculated, and non-tender. It is an uncommon entity with distinct clinical and histopathological features. Awareness about this lesion, its clinical presentation, morphological features, and favourable outcome is necessary so that it can be differentiated from other skin neoplasms.

Keywords: Dermal adipocytes, hamartoma, skin neoplasms

Introduction

Nevus lipomatosus superficialis (NLS) is an unusual, benign, cutaneous hamartomatous lesion presenting with yellowish or skin-coloured soft nodules or papules having either smooth or wrinkled surface which show mature adipose tissue amidst the bundles of collagen in the dermis on histopathological examination.

It is categorised into two types based on clinical presentation: classical and solitary. The classical form usually localizes to the gluteal region, pelvic or upper thigh region, and lower back area. They can be present at birth or may develop in the patients at or before 20 years of age. There is a zonal pattern formed by multiple, non-tender, soft, pedunculated, skin-coloured or yellow nodules, papules, or plaques. The solitary form presents as a single solitary papule, which may be widely localised anywhere in skin. It is usually seen after the age of 20 years. Cases of NLS have no sexual predilection, similar family history, or association with congenital abnormalities.\[1\]

NLS commonly shows a decrease in the number of adnexal structures but with normal morphology. Sometimes pilar abnormalities like hypertrophic pilosebaceous units, perifollicular fibrosis, abortive hair germ-like structures, fibrofolliculomas, and folliculosebaceous cystic hamartomas (FSCHs) can, however, be identified.\[1\]

We present a rare case of an 11-year-old child presenting with a swelling since birth in the lower back region.

Case Report

Patient information and clinical findings

An 11-year-old male child presented to the clinic with asymptomatic painless swelling measuring 4.8 x 3.6 cm in the lower back region. The swelling was present since birth and gradually increased in size over the past six months with no history of any neurological deficit. Examination revealed single skin-coloured swelling, which is lobulated, pedunculated, soft, and non-tender with a smooth surface. There was no ulceration,
induration, or presence of café au lait spots. There was no regional lymphadenopathy.

**Diagnostic assessment and therapeutic intervention**

Fine needle aspiration cytology (FNAC) of the swelling using a 23 G needle was performed and showed fatty material was aspirated. Both air-dried and wet fixed slides were prepared. The air-dried smears were stained with Giemsa and wet fixed smears with Papanicolaou (PAP) stain. Smears showed fibroadipose tissue along with occasional stromal fragments [Figure 1]. Findings were suggestive of lipoma, and excision biopsy was advised for histopathological examination. The swelling was excised and sent for histopathological examination in 10% formalin solution.

Grossly, the swelling was 4.8 x 3.6 x 2 cm in size. The external surface was lobulated and painted with ink. A greyish-yellow fat was seen at the base. A greyish-white lesion measuring 3.8 x 3.5 x 1 cm was identified on the cut section [Figure 2].

Haematoxylin and Eosin stained sections examined showed a lesion in the papillary and reticular dermis composed of mature adipose tissue with no connection with underlying subcutaneous tissue. Interspersed bands of collagen bundles along with scattered sebaceous and sweat glands having normal morphology were seen. The overlying epidermis was unremarkable. No evidence of malignancy was seen [Figure 3]. Features were suggestive of a benign hamartomatous lesion, nevus lipomatosus superficialis. A clinical correlation was advised.

**Follow up and outcomes**

On follow-up for two years, there was no recurrence of the lesion.

**Discussion**

NLS is a benign hamartoma of adipocytes that was first reported by Hoffman and Zurhellein in 1921.[2] It is a rare benign malformation of the skin which is characterized by the presence of mature adipocytes in the dermis histologically. The presence of fat in the dermis is variable and can range from 10% to over 50%. It has unknown pathogenesis with several proposed theories, and is not heritable.[3]

NLS is divided into two categories, the classical type, and the solitary type. The classical one usually presents at birth or infancy and is multiple, zonal, non-tender, pedunculated, cerebriform, skin-colored papules or nodules usually along the cutaneous cleavage lines. They are generally unilateral with a predilection for sites like the pelvic area, gluteal region, abdomen, and back. However, the second one, the solitary form, has no site predilection and even occurs at unusual sites. In this, the patients present in the adult stage with small solitary nodules at any site, usually over the arms, axillae, ears, scalp, and knees.[4] Unusual localisation like clitoris, scalp, and nipple has also been reported in the literature.[5,6]

The exact pathogenesis of NLS is not established yet. One of the theories is that NLS is adipose metaplasia occurring as a degenerative change in the connective tissue of dermis and heterotopia of adipocytes, or development of mature adipocytes from primitive lipoblasts by mesenchymal perivascular cells.[7] Robson et al.[8] considered it to be a true nevus due to its localization and the morphologic changes such as hyperkeratosis, hyperpigmentation, acanthosis, and also because adipose cells appear to penetrate from the subcutaneous tissue into the dermis. Yakovlev et al.[9] recommended that angiomatosis and other connective tissue nevi had an ectodermal origin, and they were not just mesodermal maldevelopments. Considering the clinicopathology of vascular neurocutaneous syndrome, it was concluded that in the background of congenital malformation of the neuro-ectodermal tissues, there is a development of superimposed hyperplasia of mesodermal tissues.[8] Hence, neuro-ectodermal melanocytic proliferation as sheets or fascicles of nevus could be associated with hamartomatous changes of mesodermal adipocytes.[9]
Some researchers regard NLS to be a kind of connective tissue nevus because mesenchymal elements in the dermis besides fat cells, like elastic fibers, collagen bundles, fibroblasts, and blood vessels, are altered in NLS. Occasionally connective tissue hamartomas have also been reported to show altered epithelial elements. A rare case of NLS with a 2p24 deletion has been reported. Further research to identify the association of NLS with genetic abnormalities or connective tissue nevus has not yet been done.[1]

An association with café-au-lait macules, leukodermic spots, comedo-like lesions, hypertrichosis on the lesion, and angiookeratoma of Fordyce is reported in cases of NLS.[4]

Cases showing concomitant NLS and deep penetrating nevus are also reported.[9]

NLS should be differentiated from other lesions like lipofibromas, skin tags, hemangioma, lymphangioma, and focal dermal hypoplasia. Histopathologic assessment helps in differentiating NLS from other lesions. No skin appendages but fat cells in the dermis are seen in lipofibromas. Skin tag, hemangioma, and lymphangioma show an absence of fat cells. Focal dermal hypoplasia shows extreme attenuation of collagen, although it shows the presence of fat cells in the dermis.[10]

Surgical excision is the preferred treatment that is curative and done for cosmetic purposes. Cryotherapy may be used in patients not willing to undergo surgery; however, it yields partial but satisfactory results.[4]

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Conflicts of interest
There are no conflicts of interest.

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