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Case Report

Pathology Section

Solitary Nevus Lipomatosus Cutaneous Superficialis: A Case Report

KULDIP SINGH CHAHAL, SWATI, NAVPREET KAUR, MONINDER KAUR

ABSTRACT

Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare benign hamartomatous skin tumou r. It consists of infiltration of mature adipose tissue into the dermal collagen. It's clinically classified in two types: the classical form, and the solitary form. We report the case of a 36-year-old male developing a solitary skin-coloured, soft, pedunculated, asymptomatic lesion over the posterior aspect of right thigh having thick cerebriform surface progressing over 1.5 year. We report this case as it is an uncommon condition with the solitary variant being more rare and to highlight its histopathological features, and differential diagnosis.

Keywords: Cerebriform, Histopathology, Pedunculated

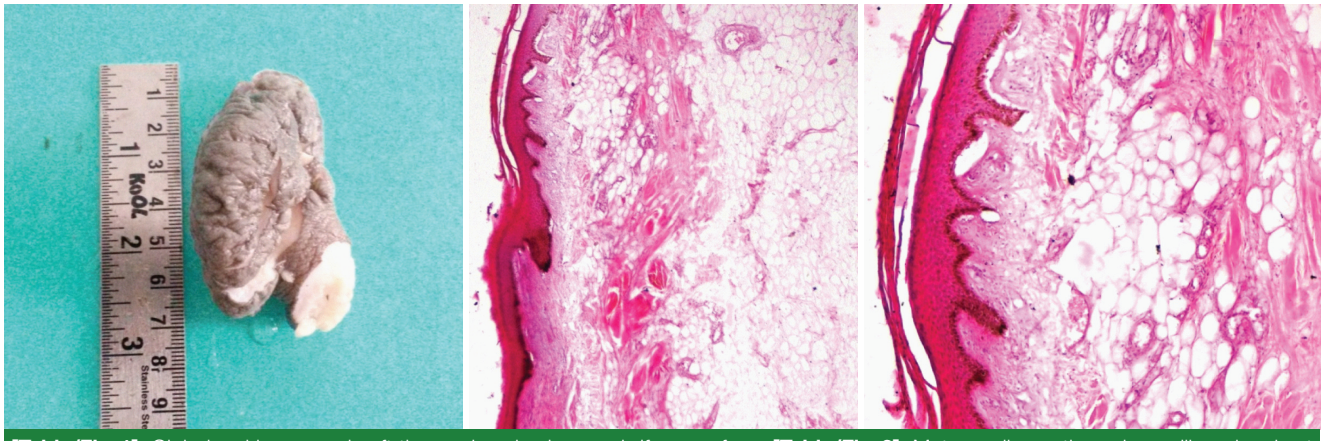
CASE REPORT

A 36-year-old male presented with a history of painless nodular swelling over posterior aspect of right thigh developing in previous 1.5 years with progression in size over 6 months. There is no history of any discharge or associated lymphadenopathy. To begin with the lesion was single and nodular over the time it progressed into a soft pedunculated lesion with thickened surface measuring 6.5X6.5X2 cm. There was no history of growth or lesion at any other site. The lesion was excised and the tissue was sent for histopathological examination after taking the informed consent from the patient.

Histopathology

Gross Examination: The specimen consisted of a skin covered pedunculated globular mass measuring 6.5X6.5X2 cm. The skin covering the lesion was thickened and wrinkled giving cerebriform appearance [Table/Fig-1], the cut surface was yellowish-white in colour.

Microscopic Examination: Microscopically the sections revealed hyperkeratosis, focal acanthosis and hyperpigmentation in the basal layer of epidermis. Vascular proliferation along with dilated ectatic blood vessels were seen in the papillary dermis. Variable amount of mature adipose tissue infiltrating the dermal collagen and around the perivascular area in the reticular and papillary dermis leading to irregular dermal subcutaneous interface was noticed. Histopathological diagnosis of Nevus Lipomatosus Cutaneous Superficialis (NLCS) was made [Table/Fig-2,3].



[Table/Fig-1]: Globular skin covered soft tissue piece having cerebriform surface. **[Table/Fig-2]:** Mature adipose tissue in papillary dermis at low power. **[Table/Fig-3]:** Elongation of rete ridges, hyperpigmented basal layer and mature adipose tissue in papillary dermis.

DISCUSSION

NLCS is a rare cutaneous hamartomatous lesion showing mature adipose tissue abnormally located in the papillary and reticular dermis [1]. Clinically the tumour can be divided into classical type (multiple) or solitary type [1]. The classical type was first reported by Hoffman and Zurhelle, usually situated on the pelvic girdle area in a segmental distribution and occurs at birth or during first three decades of life. The lesion consists of multiple, soft, non tender, pedunculated, cerebriform, yellowish or skin coloured papules or nodules [2]. The solitary type is mostly seen in adults after the age of 20 years, with no predilection for any site and may occur at unusual sites like scalp and clitoris [3-4]. Histopathologically both types consists of varying proportion of mature adipose tissue infiltration 10%-50% into the dermis [5]. No associated predilection for gender, congenital anomaly or family history is known [1] .

Clinically, there are two types: Classical (multiple) and Solitary [5,6]. The classical type presents at birth. It is mostly in the pelvic girdle, buttocks, lower back or upper thighs. Classical type is characterized by cerebriform masses or plaques consisting of numerous papules and nodules distributed a zosteri-form fashion [6]. The lesions are usually confined to one side of the midline and it grows slowly. If left untreated the nodules can coalesce and reach a huge size, so far the largest reported size is 40 × 28 cm [7].

The solitary form of NLCS usually appears during the third to sixth decades of life and can occur anywhere on the skin [8]. The nodule usually mimics a skin tag. The solitary form is also referred to as pedunculated lipofibromas [9].

The degenerative changes in dermal collagen and elastic tissue are considered as the cause of deposition of adipose tissue within the dermis [10].

The proposed pathogenesis of NLCS includes adipose metaplasia in dermal connective tissue. The developmental displacement of adipose tissue and alternatively, the origin of adipocytes from the pericytes of dermal vessels are the other proposed mechanism for its development [11].

The major differential diagnosis of NLCS include nevus sebaceous, skin tags, neurofibroma, lymphangioma, hemangioma, and focal dermal hypoplasia (Goltz syndrome), cylindroma, trichoepithelioma, and angiolipoma.

Histopathologic evaluation usually helps in the differentiation. In skin tags, no fat cells are present in the dermis. In some melanocytic nevi, pedunculated lipofibromas, and in Goltz syndrome similar features with dermal collections of adipocytes are seen on histopathologic examination. Lipofibromas contain fat cells, but skin appendages are not seen, in the dermis. In case of Goltz syndrome, the dermis is atrophic with absence of collagen and skin appendages.

A cytogenetic study on NLCS was reported recently showing a 2p24 deletion [12].

Treatment, of this condition is mostly done for cosmetic reasons. Simple surgical excision appears to be curative as the condition rarely reoccurs. No associated systemic abnormalities and malignant alterations are identified [13].

CONCLUSION

The case is reported because the solitary type of NLCS is a rare condition. It presented in the thigh region of a 36-year-old male. The clinician should be aware of this rare benign tumour and its various differential diagnosis. This tumour can attain large size if untreated.

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AUTHOR(S):

1. Dr. Kuldip Singh Chahal
2. Dr. Swati
3. Dr. Navpreet Kaur
4. Dr. Moninder Kaur

PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of Pathology, Government Medical College, Amritsar, Punjab, India.
2. Junior Resident, Department of Pathology, Government Medical College, Amritsar, Punjab, India.
3. Junior Resident, Department of Pathology, Government Medical College, Amritsar, Punjab, India.
4. Junior Resident, Department of Pathology, Government Medical College, Amritsar, Punjab, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Swati,
Flat no 20, 2nd Floor Saraswati Vihar,
Jalandhar City, Jalandhar-144008, Punjab, India.
E-mail: swatisharma13412@yahoo.in

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