Asymptomatic Firm Cerebriform Swelling Over the Lower Back

A 25-year-old male presented with asymptomatic, slowly progressive, giant swelling over the right lower back since last 4 years. There was no history of any locoregional or systemic complications. Mucocutaneous examination revealed single, well-defined, firm, protuberant, swelling of size 20 × 10 cm with very prominent cerebriform surface over the right lower back. There were discrete areas of darker pigmentation over the surface of the plaque. The plaque had a close resemblance to the brain (cerebriform) in form of very deep as well as superficial sulci and gyri [Figure 1a]. Lesion started as a single lump and gradually increased in size over the past 4 years. Systemic and other mucocutaneous examinations were within normal limits. Histopathology showed mature adipocytes in the upper, mid, and deep dermis in the perivascular and periadnexal location and between the collagen bundles [Figure 1b and c].

Question

What is the diagnosis?
Nevus lipomatosus cutaneous superficialis (NLCS).

**Discussion**

NLCS was first described by Hoffmann and Zurhelle in 1921. Clinically, it is divided into classical and solitary forms. The classic form is usually present at birth or emerges during the first two decades of life. It presents as a cluster of soft-to-firm, sessile or pedunculated, fleshy, skin-colored or yellowish nodules and plaques in the back, pelvic, and gluteal region. The solitary type usually appears during the third to sixth decades of life and can occur anywhere on the skin and have been noted in rare sites such as the scalp, eyelids, nose, and clitoris. It shows slightly different clinical and pathological features compared to classical type, and hence, few authors have referred it to as pedunculated lipofibroma.

Various atypical forms of NLCS, i.e., giant, associated with foul smelling discharge, zosteriform, with overlying hypertrichosis, and comedo-like lesion have been reported in the literature. NLCS can occur in the presence of other cutaneous conditions such as follicular papules and hypertrophic pilosebaceous lesions, angiookeratoma of Fordyce, trichofolliculoma, deep penetrating nevus, basal cell carcinoma, and hemangioma. It is presumed to be the result of displacement of subcutaneous adipose tissues embedded into the dermis but the exact pathogenesis is unknown. Electron microscopy has shown that lipocytes are closely associated with capillaries, with the suggestion of perivascular origin of young adipocytes from pericytes.

Histologically, it shows collagen bundles of the dermis intersected by mature fat cells which frequently extend into the papillary dermis. The ectopic dermal adipocytes should not be present in the continuity of subcutaneous fat tissue. This histopathological feature is considered as the most specific and necessary criterion for diagnosis. However, rarely the demarcation line between the dermis and hypodermis may be ill-defined or lost in the presence of relatively large amounts of fat in large-sized lesions. Usually, the fat cells are mature, occasionally small, incompletely lipidized, immature cells may be observed in the perivascular location. NLCS can be associated with pilar anomalies such as abortive hair germ-like structures, hypertrophic pilosebaceous units, fibrofolliculomas, perifollicular fibrosis, and folliculosebaceous cystic hamartomas.

Clinical differentials of NLCS are nevus sebaceous, neurofibroma, lymphangioma, cylindroma, trichoepithelioma, and angiolipoma. Histopathological evaluation is required to rule out these differentials. Ectopic mature adipose tissue in dermis can be seen in Goltz syndrome and large acrochordons, but they have distinctive clinical morphology.

Wide excision with skin grafting is considered as the best choice of treatment. The chance of recurrence after complete surgical excision is rare. Cryotherapy may be tried in patients who are not willing to undergo surgery. Malignant degeneration and systemic involvement are not reported in association with NLCS.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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