

Case Report

Mimicking the malignant: Solitary nevus lipomatosus cutaneous superficialis with vascular malformation

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ABSTRACT

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare, benign hamartomatous dermal lesion characterized by ectopic mature adipocytes within the dermis. Although typically asymptomatic and localized, atypical variants can pose diagnostic dilemmas. We report a 16-year-old male presenting with a gradually enlarging, bluish, non-compressible left flank lesion. Imaging suggested a vascular or soft-tissue neoplasm. Histopathological evaluation of the excised specimen revealed mature adipocytes interspersed with dilated vascular channels in the dermis, confirming NLCS with a coexisting vascular malformation. NLCS most often occurs in childhood or early adulthood, either as classical multiple plaques or a solitary nodule. Rare associations, such as vascular malformations, can mimic malignant tumors and confound diagnosis. Histopathology remains the gold standard for confirmation. This case highlights an unusual presentation of solitary NLCS with vascular malformation, underscoring the importance of histological evaluation and documentation of rare morphological variants to avoid overtreatment.

Keywords: Cutaneous hamartoma, Hamartoma, Nevus lipomatosus cutaneous superficialis, Soft-tissue tumor mimic, Vascular malformation

INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is an infrequent benign hamartomatous condition of the skin first described by Hoffmann and Zurhelle in 1921. It is histologically defined by the presence of mature adipocytes within the dermis, unrelated to subcutaneous fat. Despite its characteristic microscopic features, its clinical presentation is variable, often leading to misdiagnosis. Due to its rarity, epidemiological data are sparse, with most literature comprising case reports and small series.^[1,2]

Clinically, NLCS presents in two forms: the classical or multiple type, which appears at birth or in early childhood as grouped papules or plaques, and the solitary form, which arises later in life as isolated nodules.^[1,3] The lesions are typically asymptomatic and localized to the pelvic girdle, gluteal region, or lumbar area, though cases involving the scalp, face, and extremities have been reported.^[4,5]

Due to its benign nature, NLCS is often overlooked in the differential diagnoses of soft-tissue tumors, particularly when lesions present with atypical features. We present a case of solitary NLCS in a young adolescent male was associated with a vascular malformation—an unusual

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finding, resulting in a clinical suspicion of soft-tissue sarcoma.

CASE REPORT

A 16-year-old male presented with a 4-month history of gradually enlarging, painless swelling over the left flank. Examination revealed a soft, ill-defined, bluish, non-compressible lesion without surface changes. Contrast-enhanced computed tomography identified an ill-defined heterogeneously enhancing subcutaneous lesion ($8.5 \times 1.6 \times 5.2$ cm) localized to the lateral abdominal wall above the anterior superior iliac spine. A vascular or soft-tissue neoplasm was considered [Figure 1].

Gross pathology of the excised $10 \times 3.5 \times 1.8$ cm mass revealed multiple gray-yellow nodules beneath intact skin. Microscopy demonstrated mature adipose lobules interspersed with irregularly dilated vascular channels within the dermis. The epidermis was unremarkable. These findings were consistent with NLCS accompanied by vascular malformation [Figure 2].

DISCUSSION

NLCS represents a benign dermal malformation characterized by the ectopic presence of mature adipocytes within the dermis, without connection to the underlying subcutaneous fat. Though histologically distinctive, it remains underrecognized due to its clinical resemblance to other dermal or subcutaneous lesions.^[1]

NLCS is a rare entity, with fewer than 100 cases comprehensively described in the literature.^[1,2] It occurs sporadically, without sex predilection, and is typically seen either in early childhood (classical form) or adulthood (solitary form). The classical form presents as multiple clustered, soft, pedunculated, or sessile papules that may coalesce into plaques, often arranged in a linear or zosteriform pattern. These lesions predominantly affect the pelvic girdle, gluteal, sacral, and lumbar regions.^[1,6] In contrast, the solitary form presents as a single, skin-colored, or yellowish nodule with a broader anatomical distribution, including the scalp, face, axilla, arms, and thighs.^[3,7]

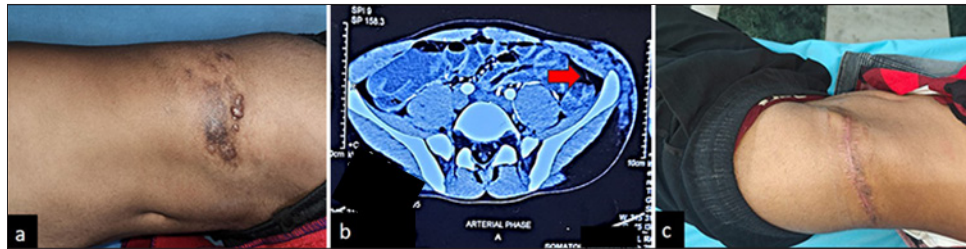


Figure 1: (a) Clinical image of soft, ill-defined, bluish, non-compressible lesion. (b) Computed tomography showed an ill-defined heterogeneously enhancing subcutaneous lesion localised to the lateral abdominal wall (red arrow). (c) Post operative image of the surgical site on the lateral abdominal wall.

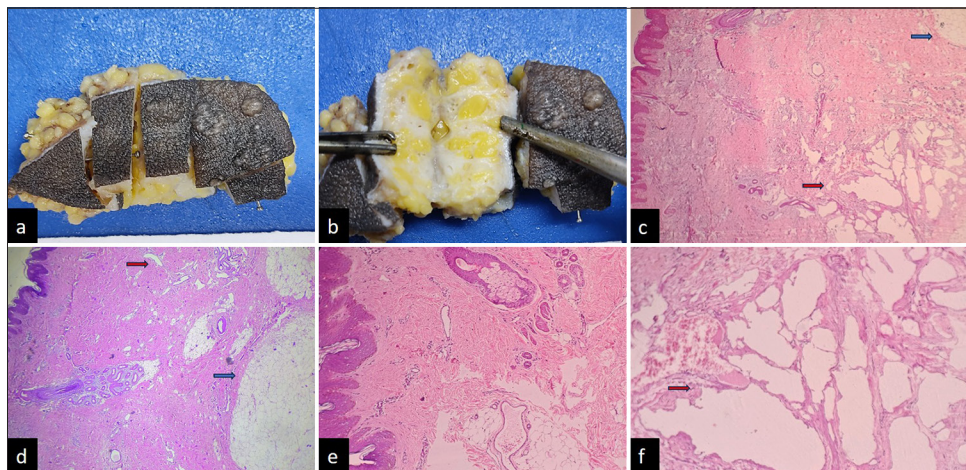


Figure 2: (a-b) Gross specimen received in the Histopathology department revealed multiple gray-yellow nodules beneath intact skin. (c-f) Microscopy demonstrated mature adipose lobules (blue arrow) interspersed with irregularly dilated vascular channels (red arrow) within the dermis. The epidermis was unremarkable. (H&E, 100x, 100x, 200x and 200x).

The pathogenesis of NLCS remains uncertain. Theories suggest adipose metaplasia of dermal connective tissue or a developmental anomaly involving perivascular mesenchymal cells or pericytes capable of adipocytic differentiation.^[1,8] Embryologically, it is speculated that NLCS arises from displaced adipose precursors or aberrant mesenchymal tissue migration. Some have proposed that the lesions may represent a form of connective tissue nevus, where dermal adipocytes accumulate due to focal mesodermal dysgenesis.^[4,6]

NLCS is usually asymptomatic. Patients may notice a slow-growing, soft dermal nodule, or plaque. Lesions may remain stable for years or slowly enlarge. In rare instances, ulceration, foul-smelling discharge, or comedo-like plugs may develop due to ischemic changes or trauma.^[5,9] Importantly, in cases like ours, overlying discoloration or vascular involvement may mislead clinicians into suspecting a neoplastic process, such as angiolipoma or sarcoma.

Microscopically, NLCS is defined by the presence of mature adipocytes within the dermis, often clustering in lobules and surrounded by normal collagen bundles. The overlying epidermis is usually unremarkable, though hyperkeratosis and acanthosis may occasionally be present.^[1,5] Ectopic adipocytes may extend to the upper dermis, sometimes with accompanying dilated vascular channels, sparse chronic inflammatory infiltrate, and attenuation of adnexal structures. In rare cases, folliculosebaceous components or cystic elements may be noted, adding to diagnostic confusion.^[5,6]

In our case, in addition to classic histological features of NLCS, we identified associated vascular malformations. This co-occurrence is highly unusual and has not been previously well-characterized in the literature. The presence of irregular, dilated vascular channels intermingled with dermal fat may confound the histopathological picture, particularly in incisional biopsies, further emphasizing the need for complete excision in ambiguous cases.

NLCS can mimic a spectrum of benign and malignant lesions. Differential diagnoses include lipomas, which are usually deeper and mobile; nevus sebaceous, which lacks dermal adipocytes; neurofibromas, which involve Schwann cells and may have a plexiform architecture; steatocystoma multiplex, characterized by multiple sebaceous cysts; angiolipomas, which have a prominent vascular component; and dermatofibromas or fibrous papules.^[3,5,8] Dermatoscopy and imaging can aid in evaluation, but definitive diagnosis hinges on histopathology.

Although benign and non-progressive, NLCS may lead to unnecessary alarm due to its resemblance to malignant lesions, especially when presenting with atypical features such as rapid growth, discoloration, or vascular involvement.^[4,6] Complete surgical excision is curative and

serves both diagnostic and cosmetic purposes. Recurrence is rare following total excision. There is no known risk of malignant transformation, and systemic associations are exceedingly rare.^[1,9]

From a clinical standpoint, the unusual presentation in our case—solitary NLCS with coexisting vascular malformation and bluish discoloration—created significant diagnostic ambiguity. The lesion's clinical resemblance to vascular tumors or sarcomas prompted cross-sectional imaging and surgical referral. This highlights the real-world impact of histological clarity in managing dermal lesions.

Documenting atypical presentations of rare entities like NLCS serves several purposes. First, it expands the spectrum of known clinicopathologic variants, equipping clinicians to recognize them earlier. Second, it underscores the utility of histopathological evaluation even in seemingly benign cutaneous swellings. Third, reporting rare associations such as vascular malformations helps build cumulative knowledge, potentially revealing underlying developmental or genetic links. Finally, these reports reinforce a multidisciplinary approach—combining clinical, radiologic, and pathologic insights—to achieve accurate diagnoses and avoid overtreatment.^[3,5,10]

CONCLUSIONS

NLCS, while benign, can exhibit atypical clinical and histological features that simulate malignant lesions. This report emphasizes the diagnostic pitfall posed by rare associations such as vascular malformations, warranting greater awareness among clinicians and pathologists.

Author's contributions: MS: Collected the data and images; NAS, TJ: Diagnosed the case on histopathology; RS, SB: Saw the patient in the clinics, treated him and performed the surgery; TJ: Wrote the manuscript. All authors reviewed and approved the manuscript.

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Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for 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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