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Case Report

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Nevus Lipomatosus Cutaneous Superficialis (NLCS): A benign cutaneous hamartoma

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Abstract

Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare, benign cutaneous hamartoma characterized by the presence of mature adipocytes ectopically located within the dermis [1]. It is considered a developmental anomaly rather than a true neoplasm. NLCS typically presents at birth or during early childhood and most commonly involves the lumbosacral region, buttocks, and thighs. Clinically, two variants are recognized: the classical form, which presents as multiple, soft, pedunculated or cerebriform papules and nodules, and the solitary form, which manifests as a single lesion [2].

We report the case of a 27-year-old female who presented with multiple, asymptomatic, skin-coloured to hyperpigmented papules on her back. The lesions were present since birth and had progressively increased in number and size. On examination, they appeared as grouped, soft, wrinkled, and pedunculated papules, suggestive of the classical form of NLCS. Histopathological examination confirmed the diagnosis, revealing polypoidal skin with focal hyperkeratosis, pigmentation of basal keratinocytes, melanin pigment incontinence, and mature adipocytes interspersed within a loose fibrocollagenous dermis containing dilated capillaries.

Differential diagnoses such as neurofibromatosis, epidermal nevus, and connective tissue nevi were excluded based on clinical morphology and histopathological findings [3]. NLCS is typically asymptomatic and does not require treatment unless there are cosmetic concerns or friction-induced irritation. Surgical excision remains the preferred treatment modality, offering excellent cosmetic outcomes with a low recurrence rate. Laser therapy may be considered for smaller lesions.

Recognizing the clinical and histological features of NLCS is crucial to avoid misdiagnosis and unnecessary interventions. Increased awareness among clinicians can lead to timely diagnosis, appropriate reassurance, and optimal management of this benign condition.

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Introduction

Nevus Lipomatosus Cutaneous Superficialis (NLCS) represents a rare, benign hamartomatous dermatosis distinguished by the presence of mature ectopic adipocytes within the dermis. NLCS has since been recognized as a cutaneous anomaly with diverse clinical morphologies, most frequently emerging either congenitally or during early childhood [1]. While the exact etiopathogenesis of NLCS remains incompletely understood, prevailing hypotheses suggest a developmental aberration resulting in focal heterotopic adipose infiltration rather than a true neoplastic transformation [2].

Clinically, NLCS is categorized into two principal variants: the classical or multiple type, characterized by grouped, soft, pedunculated or cerebriform papules and nodules; and the solitary form, presenting as an isolated lesion with a more delayed onset. These lesions are typically asymptomatic, slow-growing, and localized predominantly to the lumbosacral region, buttocks, and proximal thighs. However, atypical presentations involving other anatomical regions have also been documented [3].

The differential diagnosis encompasses a wide spectrum of dermatologic conditions, including but not limited to neurofibromatosis, epidermal nevus, and connective tissue nevi, necessitating histopathological confirmation for accurate diagnosis. Given the benign and indolent nature of NLCS, intervention is often dictated by cosmetic or functional considerations rather than medical urgency.

We herein report the case of a 27-year-old female with congenital classical NLCS involving the upper back, emphasizing its clinical and histopathological features, and highlighting the importance of distinguishing it from other cutaneous hamartomas and nevoid conditions.

Case report

A 27-year-old female presented to our dermatology outpatient department with a long-standing history of multiple cutaneous growths over the upper back. The lesions were congenital, noted at birth, and had gradually increased in both size and number over the ensuing years. The patient denied any associated pruritus, pain, discharge, or trauma to the area. There was no history of similar lesions in the family, and her medical history was otherwise unremarkable.

On clinical examination, grouped, soft, pedunculated, and wrinkled papules and nodules were observed over the right scapular region. The lesions were skin-colored to hyperpigmented, non-tender, and compressible, ranging from 0.5 to 1.5 cm in diameter. The surface exhibited a cerebriform texture in certain areas, and the lesions coalesced to form a plaque-like configuration with no signs of ulceration or secondary changes (Figure 1). The distribution and morphology were consistent with the classical variant of NLCS.

Differential diagnoses considered at this stage included neurofibromatosis type 1 (due to the multiplicity and soft consistency of the lesions), epidermal nevus, and connective tissue nevi. However, the absence of café-au-lait macules, axillary freckling, or Lisch nodules clinically excluded neurofibromatosis. Epidermal nevus and connective tissue nevi were considered less likely based on lesion morphology and consistency.

A skin biopsy was performed for histopathological evaluation. Hematoxylin and eosin-stained sections revealed polypoidal epidermis with focal orthokeratosis and basal layer hyperpigmentation. There was pigment incontinence noted in the superficial dermis. Most notably, the dermis exhibited a loose fibrocollagenous stroma with dilated capillaries and islands of mature adipocytes dispersed between collagen bundles, extending into the upper and mid-dermis (Figures 2 & 3). No evidence of neural proliferation, increased fibroblasts, or mucin deposition was noted.

These histopathological findings were diagnostic of nevus lipomatosus cutaneous superficialis, effectively ruling out the aforementioned differential diagnoses. A final diagnosis of congenital classical NLCS was established.

The patient was counselled regarding the benign nature of the condition. Given the absence of symptoms and the patient's lack of cosmetic concern, conservative management was adopted. The option of surgical excision was discussed should the patient desire cosmetic correction in the future.

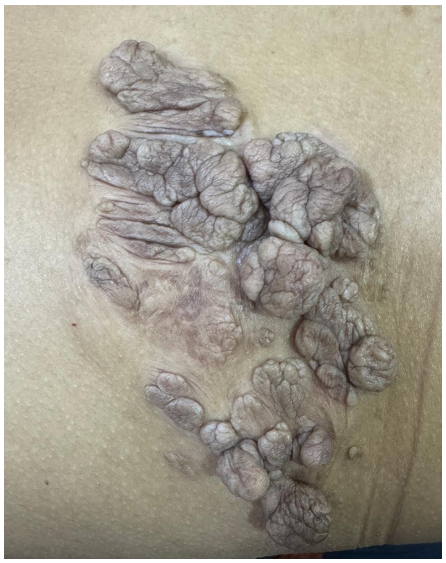

Figure 1: Grouped, soft, wrinkled, and pedunculated papules, consistent with the classical variant of NLCS.

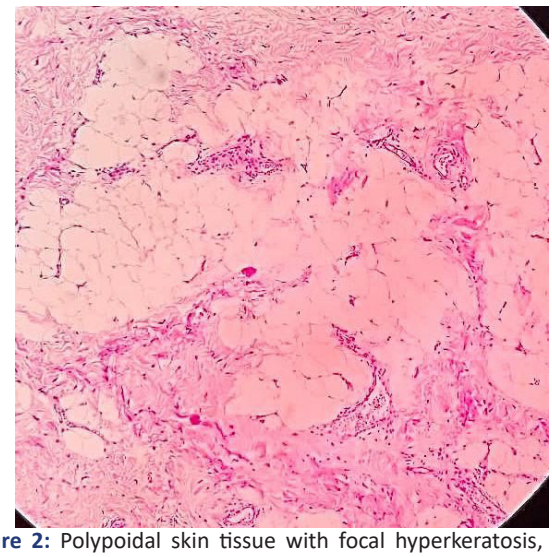

Figure 2: Polypoidal skin tissue with focal hyperkeratosis, basal keratinocyte pigmentation, and melanin pigment incontinence.

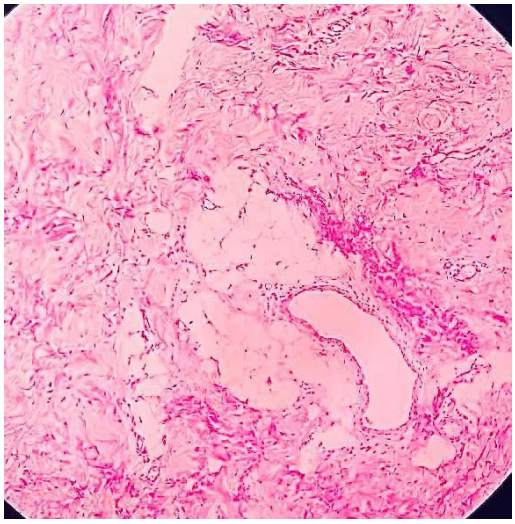

Figure 3: Loose fibrocollagenous stroma with dilated capillaries and interspersed mature adipose tissue strands.

Discussion

NLCS is an infrequently encountered cutaneous hamartoma with distinct clinical and histopathological characteristics. The classical form typically manifests within the first two decades of life and demonstrates a predilection for the pelvic girdle and adjacent areas, with congenital cases often noted at birth. However, in our patient, the lesions were first noticed at birth, supporting the congenital presentation of NLCS. The slow and insidious progression of the lesions, as well as their tendency to increase in both size and number over time, further supports the benign, non-neoplastic nature of the condition.

The pathogenesis of NLCS remains largely speculative. It is widely believed to be a developmental anomaly, with theories suggesting that it results from aberrant adipocyte differentiation during dermal development. However, no definitive genetic or molecular mechanisms have been identified to date. The classical form, as seen in our patient, is characterized by multiple, soft, yellowish to hyperpigmented papules or nodules, often involving the lumbosacral region, buttocks, or thighs. Our case involved the upper back, an atypical location but consistent with the generalized distribution reported in some cases.

Histologically, NLCS is distinguished by the presence of mature adipocytes embedded within a loose fibrocollagenous dermal stroma. The distribution of adipocytes is often in clusters or strands, typically extending into the upper and mid-dermis. In our patient, the adipocytes were surrounded by dilated capillaries and a fibrocollagenous stroma, findings that are characteristic of this condition and help differentiate NLCS from other dermatologic entities in the differential diagnosis, such as neurofibromatosis, epidermal nevi, and connective tissue nevi.

The differential diagnosis of NLCS is broad and includes neurofibromatosis type 1, which presents with multiple neurofibromas, café-au-lait spots, and Lisch nodules, and epidermal nevus, which manifests as linear or whorled hyperkeratotic lesions. Additionally, connective tissue nevi, which typically present as firm, skin-colored papules or plaques, must also be considered. However, the absence of systemic manifestations and the unique histopathological features of NLCS allowed for a definitive diagnosis in our patient.

Management of NLCS is largely conservative, given its benign and asymptomatic nature. Surgical excision is typically performed for cosmetic reasons, and the recurrence rate after excision is generally low. Laser therapy may be considered for smaller lesions, although there is limited literature on its efficacy. Our patient did not express concern regarding the cosmetic appearance of the lesions, and thus, no immediate intervention was pursued.

Conclusion

Nevus lipomatosus cutaneous superficialis is a rare, benign, and often congenital hamartomatous dermatosis that presents with characteristic soft, pedunculated lesions, most commonly affecting the lumbosacral region. Histopathological examination is essential for distinguishing NLCS from other cutaneous entities with similar clinical features. Management is primarily conservative, with surgical excision reserved for cases where cosmetic concerns arise. Patient education and reassurance are key in the management of NLCS, as the condition generally does not affect systemic health or require urgent intervention.

Conflict of interest: The authors declare no conflict of interest.

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