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CLINICAL CASE

Clinical and dermoscopic findings of classic-type nevus lipomatosus cutaneous superficialis in an adolescent. Case report

Juan A. Godínez-Chaparro[1], Marissa de J. Quintal-Ramírez[2], and Baldomero García-Gálvez[3]*

[1]Pediatric Dermatology Service, High Specialty Medical Unit of the Dr. Gaudencio González Garza General Hospital, Centro Médico Nacional la Raza, Instituto Mexicano del Seguro Social (IMSS); [2]Department of Pathology, Centro Médico Nacional la Raza, IMSS; [3]National School of Medicine and Homeopathy, Instituto Politecnico Nacional. Mexico City, Mexico

Abstract

Background: *Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a rare benign tumor characterized by the presence of ectopic adipocytes distributed among the collagen bundles in the dermis. Dermoscopy of this lesion has been poorly documented, but it aids in diagnosis.* **Clinical case:** *A female patient, 11 years and 5 months old, presented with lesions in the lumbar region that had gradually increased in size and number since birth. In September 2023, she was evaluated at CMN La Raza, where a lesion in the lumbar region was observed, consisting of a linear plaque measuring 30 × 15 mm in diameter, composed of multiple skin-colored papules and nodules that coalesced. Dermoscopy revealed a cerebriform surface, a web-like pigment network, and the presence of yellowish structureless areas. Histopathology showed a proliferation of mature adipocytes in the reticular and papillary dermis. The adipocytes were located perianexally, perivascularly, and dispersed among the collagen bundles. An increase in the density of collagen bundles was observed. The diagnosis was NLCS. The lesion was completely excised without recurrence.* **Conclusions:** *The condition may be underdiagnosed due to a lack of medical familiarity. Dermoscopy facilitates a more accurate diagnosis. An increased number of reports will contribute to the creation of a database for future studies and potential associations.*

Keywords: *Nevus lipomatosus cutaneous superficialis. Nevus lipomatosus superficialis. Hamartoma.*

Hallazgos clínicos y dermatoscópicos del nevus lipomatoso cutáneo superficial de tipo clásico en un adolescente. Reporte de caso

Resumen

Introducción: *El nevo lipomatoso cutáneo superficial es una lesión tumoral benigna rara caracterizada por la presencia de adipocitos ectópicos distribuidos entre los haces de colágeno dérmico. La dermatoscopia de esta lesión ha sido poco documentada y apoya en su diagnóstico.* **Caso clínico:** *Paciente de sexo femenino de 11 años y 5 meses de edad. Desde su nacimiento presenta lesiones en la región lumbar que han aumentado de tamaño y número de forma gradual. En septiembre de 2023 se revisó en el Centro Médico Nacional La Raza y se observó una lesión en región lumbar constituida por una placa de distribución lineal de 30 × 15 mm de diámetro, compuesta por múltiples pápulas y nódulos del color de la piel que se fusionaban entre sí. En la dermatoscopia se observó la superficie cerebriforme, red de pigmento en forma de telaraña y la presencia de áreas amarillentas sin estructura. La histopatología reveló en la dermis reticular y papilar una*

***Correspondence:**
Juan A. Godínez-Chaparro
E-mail: alberto.godinezch@gmail.com

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proliferación de adipocitos maduros. Los adipocitos se localizaron a nivel perianexial, perivascular y dispersos entre los haces de colágeno. Se observó un incremento en la densidad de los haces de colágeno. Se diagnosticó un nevo lipomatoso cutáneo superficial. Se resecó totalmente la lesión sin mostrar recidiva. **Conclusiones:** *La entidad podría estar subdiagnosticada por falta de familiaridad médica. La dermatoscopia facilita un diagnóstico más preciso. Un mayor número de reportes contribuirá a crear una base de datos para futuros estudios y posibles asociaciones.*

Palabras clave: *Nevo lipomatoso cutáneo superficial. Nevos lipomatosos superficiales. Hamartoma.*

Introduction

A hamartoma is a developmental anomaly involving an excess of one or more mature or nearly mature tissue structures, usually located at the site in question and often with a predominant component. Cutaneous hamartomas are abnormalities resulting from irregularities during embryogenesis or abnormal development of skin components. Although present from birth, they become more evident over time. Depending on the affected element, cutaneous hamartomas can be classified as epidermal, connective, lipomatous, adnexal, or angiomatous[1].

Nevus Lipomatosus Cutaneous Superficialis (NLCS) is a hamartoma of connective tissue characterized by clusters of mature ectopic adipocytes situated among the collagen bundles in the dermis[2,3]. It is a rare benign tumor; as of December 2023, fewer than 200 cases had been documented in the PubMed database. In addition, case reports in the Mexican population are scarce[2]. Although the initial clinical evaluation may suggest the diagnosis, dermoscopy – a diagnostic tool that has been minimally described in this type of hamartoma – provides valuable additional support in its identification. Here, we present a classic case of NLCS in a Mexican adolescent, where dermoscopy was employed as part of the diagnostic process.

Clinical case

This is a case of an 11-year-and-5-month-old female patient with a 2-year history of anxiety disorder treated with sertraline. She has no relevant medical history concerning her present condition. Since birth, she has developed lesions in the lumbar region, initially in small quantities. Over the years, these lesions have gradually increased in size and number, with very slow growth. The lesion remained asymptomatic throughout. She did not receive any medical intervention. In September 2023, she was referred to the dermatology department at CMN La Raza for evaluation.

During somatometry, the following data were recorded: Weight of 53 kg, height of 1.51 m, heart rate of 82 bpm, and respiratory rate of 21 rpm. During the physical examination, a localized dermatosis was observed on the trunk, affecting the lumbar region. The lesion consisted of a slightly oval linear plaque measuring 30 × 15 mm in diameter, composed of multiple skin-colored papules and nodules that coalesced. Dermoscopy revealed a cerebriform surface formed by gyri and sulci, with some areas showing a web-like pigment network and the presence of yellowish structureless areas, some of which displayed perifollicular distribution (Fig. 1). A skin biopsy was performed, revealing epidermal acanthosis and, in some areas, elongation of the dermal papillae. In the reticular and papillary dermis, a proliferation of mature adipocytes was observed. The adipocytes were located perianexally, perivascularly, and dispersed among the collagen bundles. An increase in the density of the collagen bundles was also noted (Fig. 2). Based on all histological findings, a diagnosis of NLCS was made. Subsequently, the lesion was completely excised by the plastic and reconstructive surgery department. To date, no recurrence of the lesion has been reported.

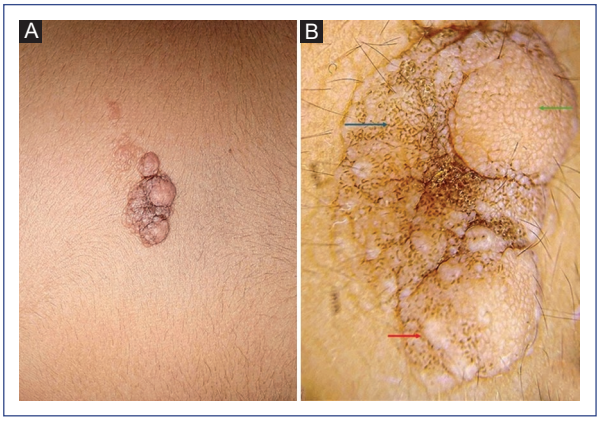

Figure 1. A: clinical image showing a linear skin-colored plaque with slight central hyperpigmentation, composed of multiple papules that group and fuse together. **B:** dermoscopy (Dermlite DL3, ×10, contact/non-polarized mode). The cerebriform surface (green arrow), some areas with a spiderweb-like pigment network (blue arrow), and the presence of structureless yellowish areas (red arrow) are observed.

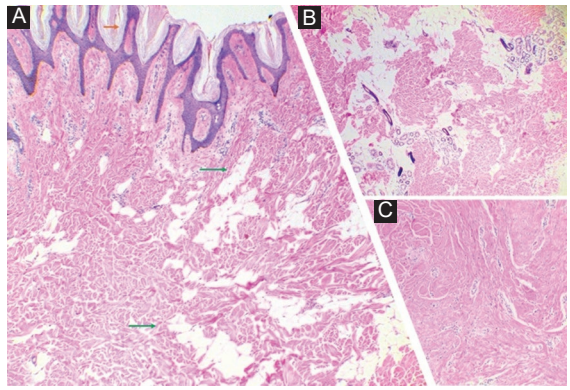

Figure 2. Histopathologic examination. **A:** panoramic image showing epidermal acanthosis and elongation of dermal papillae (orange arrow). In the papillary and reticular dermis, there is a proliferation of mature adipocytes scattered among the collagen bundles (green arrow). **B:** adipocytes are located periannexally, perivascularly, and scattered among the collagen bundles. **C:** increase in the density of the collagen bundles.

Discussion

NLCS, first described by Hoffmann and Zurhelle in 1921, has an uncertain etiology with several proposed theories. Hoffmann and Zurhelle suggested that fat deposition in the dermis is secondary to degenerative changes (metaplasia) in connective tissue[4]. Another hypothesis proposes that abnormal fat formation in the dermis might originate from perivascular precursor cells, similar to the development of fetal fat. This theory is supported by microscopic studies that have identified young adipocytes around blood vessels, which subsequently mature into fat[5]. In addition, some believe that the adipocytes represent a true nevus arising from focal heterotopic development of adipose tissue[6].

NLCS presents two distinct clinical forms: The multiple or classic type, and the solitary form. In the classic type, lesions are present from birth, although they may not be apparent during the 1st few years of life, and they progressively develop over the following three decades. These lesions consist of multiple papules that cluster to form skin-colored or yellowish plaques. They may have a linear zosteriform distribution (as in the present case) or a segmental distribution, with a smooth or cerebriform surface. Their growth is slow, but they can reach considerable sizes if left untreated and are often noticed by the patient after years. The most common locations for the classic type include the pelvic girdle (40%), the lower trunk (31%), and the lower extremities (15.5%). In contrast, the solitary form is characterized by a single sessile dome-shaped papule or nodule, typically appearing between the third and sixth decades of life, measuring 0.3-2.5 cm, and primarily located on the legs (29.4%), trunk (23.5%), and buttocks (20.5%), though it may also occur in other areas, including the scalp, eyelids, nose, and clitoris. Cases of coexistence of both clinical types in the same patient have been reported[2,3,7].

Dermoscopy is a diagnostic tool that, using an instrument combining a light source and a magnifying lens, allows the clinician to visualize subcutaneous structures that are not visible to the naked eye. Its use in non-melanocytic lesions is relatively recent, and reports on the dermoscopic features of NLCS are limited. Vinay et al. identified five dermoscopic findings in NLCS: (1) Cerebriform surface formed by gyri (ridges) and sulci (fissures) filled with keratin, (2) web-like pigment network, composed of brown lines and yellowish holes creating a honeycomb-like pattern, (3) border of the cerebriform surface with a homogeneous white film resembling “frosted glass” or a “veil,” (4) yellowish structureless areas, some with perifollicular distribution, corresponding to dermal adipocytes, and (5) comedo-like openings[8].

Baraldi et al. documented additional dermoscopic findings in NLCS, including a pink background and white linear structures. In lesions with a smooth surface, white linear structures with linear spiral vessels were observed, while in NLCS with a cerebriform surface, brown pigmented areas with irregularly distributed linear spiral and looped vessels were identified[7]. The diversity in findings highlights the need to continue describing the dermoscopic characteristics of NLCS to establish clear diagnostic patterns.

Histopathologically, the characteristic finding of NLCS is the proliferation of mature adipocytes in the reticular dermis, which may extend into the papillary dermis. Generally, there is no connection with the subcutaneous tissue; for some authors, this finding is necessary to establish the diagnosis of NLCS. The proportion of adipose tissue embedded among the dermal collagen bundles ranges from 40% to 70%, with larger lesions showing a higher quantity of adipocytes compared to smaller ones. Adipocytes are typically located periannexially (around the eccrine glands), perivascularly, or scattered among the collagen bundles. A small amount of fat may be found around subpapillary blood vessels, with a central large blood vessel surrounded by several smaller vessels giving a glomeruloid appearance. In

some cases, there is an increase in the density of collagen bundles, fibroblasts, and blood vessels within the dermis[3,4,9].

In rare instances, the coexistence of NLCS with other lesions, such as trichofolliculoma, folliculosebaceous hamartoma, or perifollicular fibroma has been observed. These hamartomas can clinically resemble other types of cutaneous tumors or hamartomas. Differential diagnosis for the classic type of NLCS includes connective tissue nevus, mucinous sebaceous nevus, pilocytic hemangioma, lymphangioma, and focal dermal hyperplasia (Goltz syndrome). The solitary type may be confused with soft fibromas, solitary neurofibromas, and solitary lipofibromas[2,3]. For an accurate differential diagnosis, the previously described dermoscopy is used, and a histopathological study is essential.

Conclusions

This condition may be underdiagnosed due to a lack of familiarity among physicians. Dermoscopy serves as a non-invasive tool in the diagnosis of NLCS and helps differentiate it from other skin conditions. In addition, documenting a greater number of cases will contribute to creating a database for future studies and potential associations.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors obtained parental informed consent and approval from the Ethics and Research Committee for the analysis of routinely collected and anonymized clinical data.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

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