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## NEVUS LIPOMATOSUS SUPERFICIALIS IN SACROCOCCYGEAL REGION. CASE REPORT IN AN INFANT

**Palabras clave:** Nevo; Lipoma; Hamartoma; Lipomatosis; Neoplasias cutáneas.

**Keywords:** Nevus, Lipoma, Hamartoma, Lipomatosis, Skin Neoplasms.

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## ABSTRACT

Nevus lipomatosus superficialis is a benign tumor characterized by the presence of mature adipocytes located in the superficial layer of the dermis; its management is surgical and most of the time is reported in adults.

This case presents a malformation from birth in the sacroccocygeal region over the lateral line to the right, which is why, after obtaining imaging studies to rule out an associated hidden dysraphism, surgery was decided for total resection of the mass, without complications. Finally, the pathology report diagnoses nevus lipomatosus superficialis, which constitutes a rare and unusual case based on the differential diagnoses and presentation.

## INTRODUCTION

Nevus lipomatosus superficialis is a benign lesion characterized by the presence of mature adipocytes in unencapsulated subcutaneous tissue (1,2), which is relevant when differentiating this lesion from lipoma.(2). Some authors consider it as a subtype of hamartoma, since its proliferation is associated with the presence of blood vessels in the dermis (3).

This lesion was first described in 1921 by Hoffman & Zurhelle as an adipocyte malformation that appeared in several clustered nodes (3), which could have a zosteriform arrangement. In 1950, Nikolowsky described a variant of the classical form (3) as a single pedunculated nodular lesion, also known as lipofriboma (3). This paper presents the case of a patient who developed the second type of the lesion; since this is rarely described in children, a histological examination of the resected mass was necessary to differentiate it from other similar lesions such as lipomas and focal dermal hipoplasia, among others.

## CLINICAL CASE

Female patient, 29 days old, from Bogotá, referred by her pediatrician due to a pedicle lesion in the sacroccocygeal region and no other history of importance. Since the lesion appeared in the midline, an MRI in the spine was performed, which discarded the presence of hidden dysraphism (Figure 1). During pregnancy, prenatal ultrasounds were normal, and the mother of the patient had no complications and did not require additional drug treatment; at the time of assessment, the mass had increased in size, but did not generate associated symptoms.

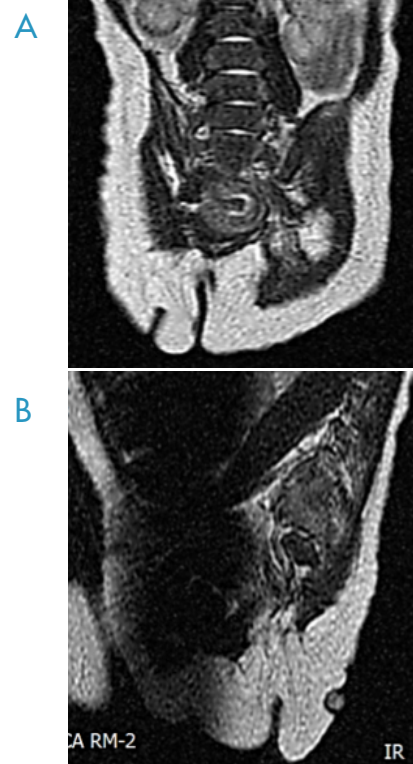


Fig 1. MRI of the lumbosacral spine.

A. Sagittal section: right paracoccygeal mass with broad based pedicle.

B. Coronal section: coccygeal region mass not connected to spinal cord

Source: Own elaboration based on the data obtained in the study.

Physical examination showed a well-defined mass in the previously described region, painless, without alterations in color, without renitency point or active secretion (Figure 2). A resection was scheduled in order to continue with histopathological studies.



Fig 2. Mass of the patient at the time of the first consultation.

Source: Own elaboration based on the data obtained in the study.

The procedure was conducted two weeks after finding the unencapsulated, non-vascularized lipomatous mass. Since it presented a broad based pedicle, a skin plasty with simple skin flaps was necessary after completing the resection procedure; the mass was sent

to pathology, which reported a mature adipose tissue of lobular appearance that completely occupied and expanded the superficial dermis with some fibrocollagenous interstitial tissue bands. The diagnosis of nevus lipomatosis superficialis or Hoffmann-Zurhelle was made based on these characteristics (Figure 3). Postoperatively, the patient had adequate cicatrization and no complications (Figure 4); during a follow-up at five months, no relapse or other alterations associated with surgery were reported.

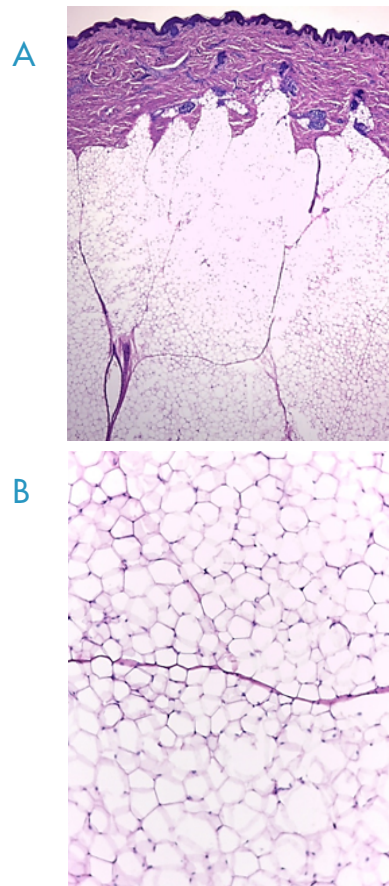


Fig 3. Histology of the resected mass.

A. Multiple partitioned lobes of lipomatous cells in epidermis.

B. Collagen band which groups lipomatous cells.

Source: Own elaboration based on the data obtained in the study.



Fig 4. Tenth day after resection of the mass.

Source: Own elaboration based on the data obtained in the study.

## DISCUSSION

The onset at birth of nevus lipomatosus superficialis and its location were noteworthy since, usually, they are a sign of hidden dysraphism in newborns (4); given these specifications, using diagnostic imaging was necessary. The characteristics of the mass allowed a simple surgical approach and resection; it is important to highlight the size of the mass. Regarding the patient, a skin plasty had to be used to provide the best possible healing.

In general, lipomatous lesions in children are rare and their incidence is less than 10% (3), while adults may show more than 50% (3); among these lesions, nevus lipomatosus superficialis represents 2% (3). The literature includes few reports of this entity not only because of its low incidence, but because it is underdiagnosed.

The pedicled lipofibroma differs from the classical form described by Hoffmann & Zurchelle (3), which may appear at any site of the body and is unique, while histological features

remain the same (5). Differential diagnoses include focal dermal hypoplasia that has a distribution of collagen beams different from that of the lipomatous nevus, is horizontal simulating a scar and presents with other congenital malformations (5). Segmental neurofibromatosis, which has similar lesions associated with light brown spots, is also included (2).

Another differential diagnosis is hidden spinal dysraphism, which causes defects in the fusion of the neural tube and causes meningeal and spinal deformations, which lead to a progressive neurological deficit that can become severe (4). Along with several entities such as tethered cord syndrome, diastematomyelia, meningoceles and myelomeningocele (4), this condition is characterized by a cover skin lesion and associated skin disorders in the sacrococcygeal region on the midline, including dimples and dermal sinuses, connective nevi, localized hypertrichosis, hemangiomas, dyschromic lesions, lipomas, among others (4). Based on these characteristics, this is the first differential diagnosis to consider when facing malformations in this location.

In conclusion, nevus lipomatosus superficialis has a low incidence in the pediatric population, is poorly described and its diagnosis can be only made through histopathology; treatment is surgical and consists of total resection. In this case, the unique pediculated variant was presented, which is less frequent than the classic one, in which, due to its location, other pathologies associated with the nervous system had to be considered in order to rule out serious neurological diseases. These diseases could be studied through imaging during the first weeks of the patient's life, so this report not only stands out because of the infrequency of the underlying pathology but also because of the differential diagnoses that had to be suspected.

## CONFLICTS OF INTEREST

None stated by the authors.

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