Lumbar paravertebral tumor in a newborn: congenital lipomatous nevus

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Abstract

Introduction. Skin lesions in close proximity to the lumbosacral region should be assessed in newborns, since they may be the first sign of hidden spinal dysraphism.

Clinical case. We present the case of a newborn without significant prenatal history. On the first day of life, a 1 cm diameter nodular lesion was found at the lumbar level of the right paravertebral region, with a vascular stain surrounding the base of the lesion. Neurological examination was normal. A soft tissue ultrasonography was carried out. It showed no continuity with the lumbar spinal canal. As a result of the lesion increasingly growing, resection was decided upon six months later. Pathological examination confirmed diagnosis – Hoffmann-Zurhelle nevus.

Discussion. Hoffmann-Zurhelle nevus is an infrequent cutaneous hamartomatous lesion in newborns. Treatment is always surgical in order to avoid potential growth-related complications.

Key Words: Newborn; Hidden spina bifida; Lipoma.

Tumoración paravertebral lumbar en el recién nacido: nevus lipomatoso congénito

Resumen

Introducción. Las lesiones en la piel de los recién nacidos situadas en la proximidad de la región lumbosacra deben estudiarse, ya que, en ocasiones son la primera manifestación de un disrafismo espinal oculto.

Caso clínico. Recién nacido sin antecedentes prenatales de interés. En su primer día de vida se observa una lesión nodular de un centímetro de diámetro situada en la región paravertebral derecha a nivel lumbar con una mancha vascular envolviendo la base de la lesión. Examen neurológico normal. Se realiza una ecografía de partes blandas donde no se evidencia continuidad con el canal medular lumbar. Seis meses más tarde debido al crecimiento progresivo de la lesión se decide su resección. La anatomía patológica confirma su diagnóstico: nevus de Hoffmann-Zurhelle.

Comentarios. El nevus de Hoffmann-Zurhelle es una lesión cutánea hamartomatosa infrecuente en los recién nacidos. El tratamiento es siempre quirúrgico para evitar posibles complicaciones relacionadas con su crecimiento.

Palabras Clave: Recién nacido; Espina bífida oculta; Lipoma.

INTRODUCTION

Spinal dysraphism refers to all malformations caused by neural tube development and/or closure disorders. Hidden dysraphism occurs when such disorders are covered by the skin, which prevents the direct exposure of the nerve tissue from being observed, thus delaying diagnosis[1].

Various cutaneous disorders associated with hidden spinal dysraphism (aplasia cutis, dermal dimple, connective nevus, dyschromia, localized hypertrichosis, neurofibroma, melanocytic nevus, lipoma, vascular lesions...) have been described. Therefore, when observed in clinical practice, additional tests are required to confirm or rule out the presence of potential malformations of the underlying backbone[2].

CLINICAL CASE

We present the case of a 39-week-old – gestational age – full-term newborn, with no significant obstetric history except for left renal agenesis found at prenatal ultrasonography. At physical exploration at the maternity department, a right lumbar paravertebral cutaneous appendage was found, with a vascular lesion at its base (Fig. 1). An abdominal ultrasonography was performed, and the absence of the left kidney was confirmed. A soft tissue ultrasonography of the lumbosacral region was conducted. It revealed no signs of spinal dysraphism, as well

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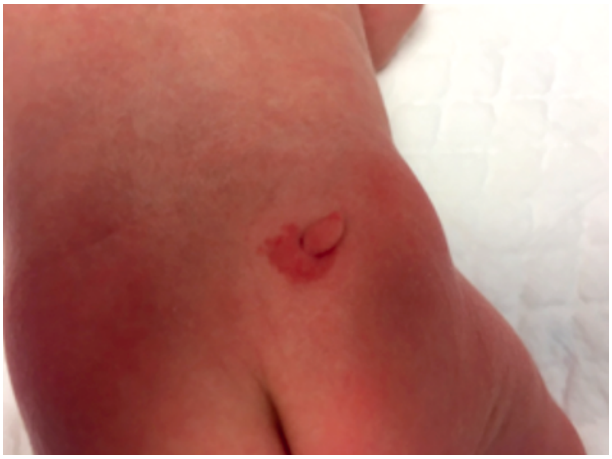

Figure 1. Cutaneous appendage above a vascular lesion in a newborn at 24 hours of life.

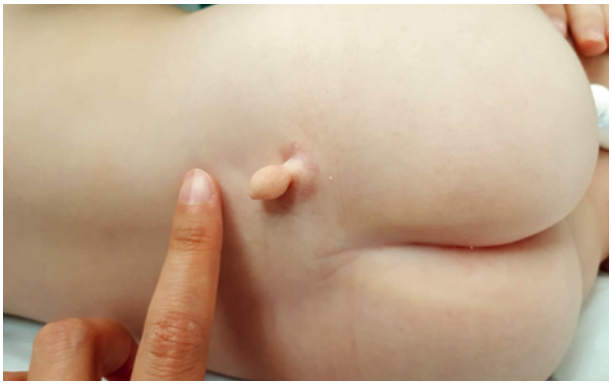

Figure 2. 4 cm diameter polypoid lesion at 6 months of life.

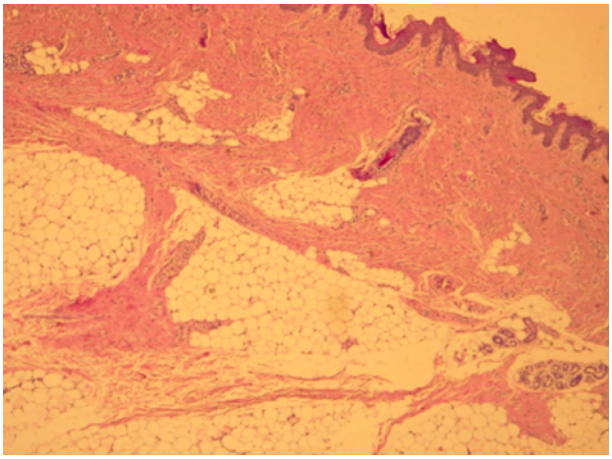

Figure 3. Adipose tissue infiltrates.

as a visible spinal cord with horsetail roots and a thecal sac compatible with normality. The lack of spinal canal disorders was confirmed by means of a lumbosacral nuclear magnetic resonance. The newborn was discharged, with subsequent outpatient pediatric follow-up.

During follow-up, the lesion progressively grew up to 4 cm of diameter (Fig. 2). The patient had normal neurological development.

As a result of malformation growing after 6 months, surgical resection was decided upon. Pathological examination of the sample revealed the presence of a skin polyp covered by an undulated epidermis, with a slight acanthosis and papillomatosis, and a connective axis made up of mature, vascularized adipose tissue lobes, compatible with superficial cutaneous lipomatous nevus or Hoffman-Zurhelle nevus (Fig. 3). Postoperative outcome was satisfactory, with adequate healing and no surgical wound superinfection.

DISCUSSION

Superficial cutaneous lipomatous nevus or Hoffman-Zurhelle nevus is a rare entity[3]. The first case was described in 1921 by Hoffman and Zurhelle, after whom it was named[4]. Today, very few cases in newborns have been described in the literature, since it mostly occurs at a later stage of life, generally between the second and the third decades. In the cases described, there is a slight predominance of female patients. Etiology is unknown, with no evidence of hereditary influence[5]. It is believed to be caused by connective tissue degeneration, with subsequent accumulation of adipocytes at the site involved[6]. Clinically speaking, the main lesion manifests as a hamartomatous nevus-like malformation, with a papular, tuberous, or nodular appearance[7]. It tends to be soft, and the skin color is typically normal. This lesion may be found both isolated and in groups, with actual plates being formed. Although more rarely, papular groups in one specific region, with a distribution similar to herpes zoster, have also been reported[8]. The most common location is the glutei, followed by the pelvic girdle and the proximal portion of both upper limbs, but it may be found anywhere in the body. Growth is progressive, with no additional symptoms. Diagnosis is established following pathological examination, which means it is achieved after resection and assessment. The main histological characteristic is the presence of well-differentiated, irregularly-distributed adipose tissue at the dermal level, without reaching the subcutaneous fat. An inflammatory infiltrate may occasionally be observed, with mononuclear cells and mastocytes at the perivascular level[9]. Differential diagnosis includes lumbosacral appendage – also known as human tail and classified by various authors as an actual tail (a distal remnant of the embryonic tail) – and sacral pseudotail – adipose tissue, connective tissue, striated muscle, vessels, nerves, and skin[10]. Other pathologies to be ruled out include sacrococcygeal eversion, lipoma, fibrolipoma, sebaceous nevus,

and lipoblastomatosis[11]. It is worth noting that segmental neurofibromatosis should be ruled out in the presence of café-au-lait spots[12]. Regarding treatment, surgical resection is indicated in case of excessive growth potentially causing further problems, locations with cosmetic implications, and diagnostic uncertainty. Sometimes, owing to the size of the lesion, repairing the defect by means of grafts or skin flaps may also be required[13]. Even though some rare cases have been reported in the literature, recurrence is infrequent following surgical removal[14]. Prognosis is good, with malignization being extremely rare[15].

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