Spinal lipoma associated with congenital dermal sinus: a case report*

Lipoma espinhal associado a seio dérmico congênito: relato de caso

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Abstract Spinal lipomas are rare, accounting for 1% of all spinal tumors and being associated with occult spinal dysraphism in more than 99% of cases. Such lesions are divided into three main types, namely, lipomyelomeningoceles, intradural lipomas, and filum terminale fibrolipomas. The present report describes a case of congenital lumbosacral lipoma associated with cutaneous stigmata of the lumbar dermal sinus type.
Keywords: Occult spinal dysraphism; Congenital dermal sinus; Intradural lipoma; Ultrasonography.

Resumo Os lipomas espinhais são raros, respondendo por 1% de todos os tumores espinhais, estando associados ao disrafismo espinhal oculto em mais de 99% dos casos. Estão divididos em três tipos principais: lipomielomeningocele, lipoma intradural e fibrolipoma do filo terminal. Este relato descreve um caso de lipoma lombossacral congênito associado a estigma cutâneo do tipo seio dérmico lombar congênito.
Unitermos: Disrafismo espinhal oculto; Seio dérmico congênito; Lipoma intradural; Ultrassonografia.

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INTRODUCTION

Occult spinal dysraphisms constitute a group of dorsal conditions lying under an intact layer of dermis and epidermis. As the skin and nervous tissue originate from the ectoderm, abnormalities may simultaneously occur in both of them. Thus, association with a cutaneous stigma such as skin covered mass, hair tuft, cutaneous appendix, skin color disorder, cutaneous depression, is frequently observed[1–3]. The spectrum of occult dysraphic lesions includes lipomas, dorsal dermal sinuses, myelocystoceles and diastematomyelia. Dorsal dermal sinus is usually associated with 60% of the cases of spinal lipoma[4].

Spinal lipomas are less frequently found and may be located either outside or inside the spinal canal, or even in a combination of these locations[1–5]. The present report describes a case of lipoma located inside the spinal canal (intradural lipoma), whose suspicion and detection were facilitated by the presence of a cutaneous stigmata of congenital dorsal dermal sinus type.

CASE REPORT

A male full-term neonate born by cesarean delivery, with antenatal ultrasonography studies with no abnormality. Clinical examination detected cutaneous stigma in the form of a cutaneous depression with a small paramedian ostium covered by a thin transparent membrane with elevated borders, immediately at left from the midline, above the intergluteal fold, compatible with dorsal dermal sinus. Also, a small solid mass covered by skin was observed in the lumbosacral region (Figure 1).

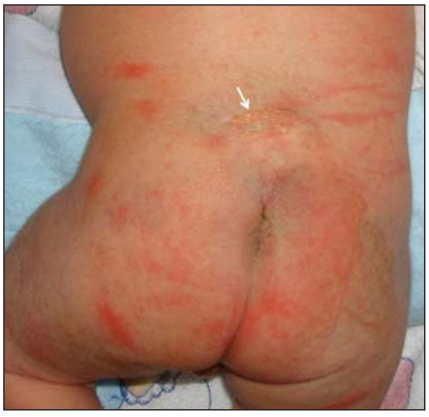

Figure 1. Dorsal dermal sinus. Image of the neonate with cutaneous stigmata (arrow) characterized by cutaneous depression covered by a thin membrane located above the intergluteal fold.

Ultrasonography of the lumbosacral spine was requested at the third day of life, upon observation of the cutaneous stigmata indicating the presence of a possible occult spinal dysraphism. The images demonstrated a hypoechoic, elongated, solid mass within the spinal canal, extending from T12 to L4, measuring 3.2 × 0.7 cm, posteriorly located and attached do the spinal bone

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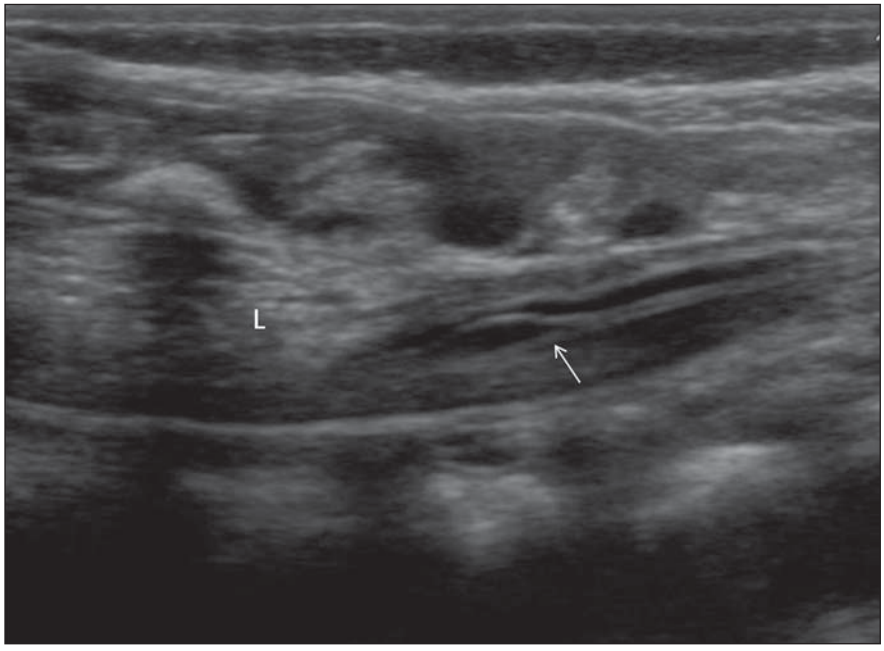

Figure 2. Tethered spinal cord syndrome. Midsagittal section at ultrasonography. The arrow indicates the medullary cone at the sacral level, as a function of the presence of the intradural lipoma (L).

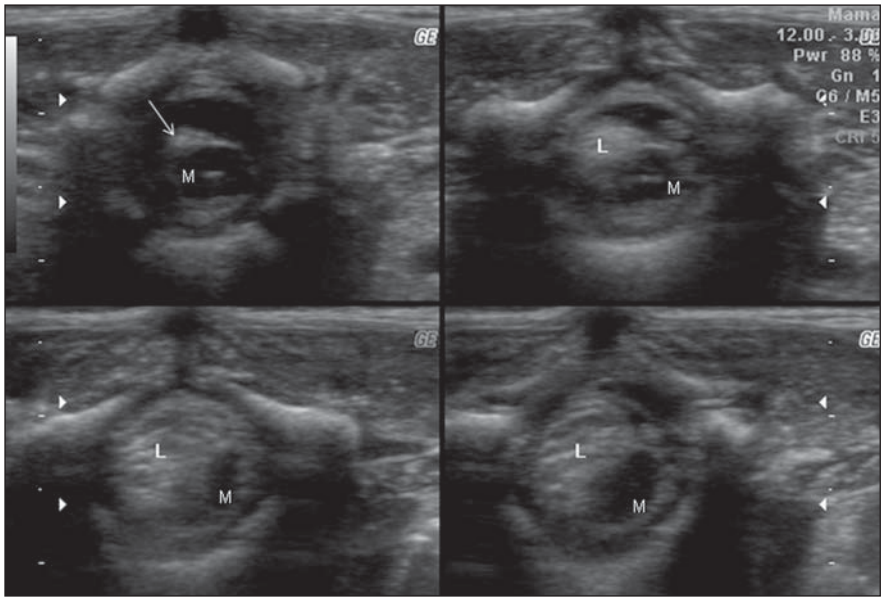

Figure 3. Cross-sectional section at ultrasonography performed with the patient in ventral decubitus, at different levels of the lumbosacral spine, demonstrating a hypoechoic, solid mass (arrow) (L), which fits into the medullary canal, attached to the bone marrow (M), displacing it anterolaterally to the right.

marrow, displacing it anterolaterally to the right and determining the presence of tethered spinal cord syndrome (Figures 2 and 3), with medullary cone at the level of the third and fourth lumbar vertebrae.

Magnetic resonance imaging confirmed the sonographic findings of a mass within the spinal canal, accurately determined the lesion topography and demonstrated the opening of the dorsal dermal sinus, from the spinal canal to the skin surface (Figure 4).

At his second month of life, the child was submitted to laminectomy with subtotal tumor resection. He remains under follow-up, with good clinical progression, and a subtle right lower limb motor deficit.

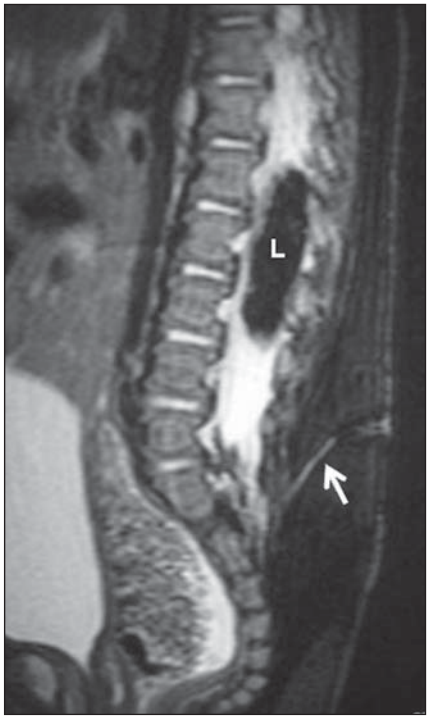

Figure 4. Mid-sagittal magnetic resonance imaging slice, T2-weighted image with fat-saturation, demonstrating intradural lipoma (L). The arrow indicates a dermal sinus connecting the fundus of the dural sac with the skin at S1–S2 plane.

DISCUSSION

Occult spina bifida is frequently asymptomatic and may clinically manifest at any age. The defect occurs at the first two months of intrauterine life. Embryologically, the neural tube develops from ectodermal cells (neuroectoderm), while the mesoderm will originate the bones, meninges and muscles. The skin is separated from the neural tube by a mesoderm layer. An incomplete separation between the cutaneous ectoderm and the neural tube results in spine tethering, diastematomyelia or in dermal sinus. The premature separation between the cutaneous ectoderm and the underlying neuroectoderm leads to incorporation of mesenchymal elements between the neural tube and the skin, which may result in the development of lipomas[6].

Spinal lipomas are divided into three main types as follows: lipomyelomeningocele, intradural lipoma and filum terminale fibrolipoma[1]. Besides fat, about 80% of such tumors may contain neural tissue or meninges[5].

Congenital dorsal dermal sinus is an epithelial connection between the skin and deepest tissues, resulting from a probable incomplete separation between the cutaneous ectoderm and the underlying neuroectoderm. Its estimated incidence in the general population is 1:2,500 live newborns. At clinical examination, it presents as a cutaneous depression or ostium, and is usually associated with 60% of spinal lipomas. The presence of such a condition should be considered as an alert sign for spinal lipoma screening by means of ultrasonography[5].

The sonographic study assumes a relevant role as a screening method in neonates under suspicion of occult dysraphism without the presence of an apparent mass. Most of times, the presence of cutaneous stigmata is the sole indicator of the possible existence of occult spina bifida. Ultrasonography presents a good sensitivity and plays a significant role in the screening for fatty masses, characterization of the medullary cone topography and in the thorough analysis of the relation between the spinal bone marrow and the possible existence of a mass.

Ultrasonography of a neonate's spinal canal plays a relevant role as screening method, but as such, it presents limitations. Most of times, this method cannot identify the communication between the dermal sinus and the dural sac, which is important to rule out the risk for meningeal infection. The method can only infer the possibility of communication. Magnetic resonance imaging is the most specific imaging method to evaluate the spinal canal in neonates, allowing diagnostic confirmation and a more detailed analysis of dysraphism[3,7].

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