Lipomyelomeningocele: A Cause of Closed Spinal Dysraphism Two Case Reports and Review of the Literature

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Abstract

Lipomyelomeningocele, a rare spinal anomaly present at birth, involves a lipoma firmly connected to the back of the neural placode, extending outward through a defect in the spine known as spina bifida and merging with subcutaneous fat tissue. MRI imaging is crucial in both diagnosing and managing this condition. Our report details the identification of a lipomyelomeningocele in two infants, aged 3 months and 18 months, respectively.

Keywords: Lipomyelomeningocele, closed dysraphism, imaging, spine.

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INTRODUCTION

Lipomyelomeningoceles, a rare form of closed spinal dysraphism or spina bifida occulta, typically manifest as a subcutaneous fatty mass located above the intergluteal cleft in the lumbosacral region. However, variations exist, and these anomalies can occur at different points along the spinal canal. This category encompasses a diverse range of malformations characterized by their enclosed nature beneath the skin [1].

Specifically, lipomyelomeningocele represents a subtype of occult spinal dysraphism where lipomatous tissue is connected to the spinal cord through a defect in the spine, incorporating the meninges or spinal cord and forming a posterior mass beneath the skin [2]. In this report, we present two cases of lipomyelomeningocele diagnosed in 3 and 18-month-old infants. Additionally, we conducted a brief literature review to recapitulate the primary imaging characteristics associated with this condition.

CASE REPORT

Case N°1

This is an 18 month old infant with a spinal deformity that progresses from birth. An x-ray of the dorsolumbar spine was performed showing dorsolumbar scoliosis. An MRI was then performed (Figure 1), showing a bony defect in the posterior arches of the lumbosacral region lateralized to the right and extending for over 4.5 cm from L4 to S1 with a large hernia of the dural sac containing cerebrospinal fluid and the nerve roots of the cauda equina with individualization within it of a subcutaneous compressive fatty formation infiltrating medially and pushing forward the right psoas muscle; appearing hyperintense in T1 weighted sequences and fading on FATSAT sequences without contrast enhancement.

This mass infiltrates the canal, attracting the roots of the cauda equina and pushing back the medullary cone, revealing syringomyelic cavities that appear hyperintense in T1 and hypointense in T2. It is associated with an expansion of the peri-medullary spaces at its level, responsible for an enlargement of the medullary canal.

The spinal CT scan, revealed dorsolumbar scoliosis associated with vertebral malformations associating a rotational component, tiered hemivertebraes from D11 to L1 and fusion of the vertebral bodies from L1 and L2.

In light of those findings, the diagnosis of lipomyelomeningocele associated with dorsolumbar scoliosis and vertebral malformations was retained.

Case N°2:

This is a 3-month-old boy admitted for management of a congenital mass in the lumbosacral region. A spinal ultrasound was performed, revealing a
dorsolumbar posterior arch defect associated with an elongated spinal cord attached to an intracanal echogenic fatty mass protruding into the lumbar subcutaneous tissue. For better characterization, a spinal MRI was conducted, (Figure 2) showing a failure of closure of the posterior arches from D11 to L5 and the sacral posterior arches. This results in a herniated sac lateralized to the left, measuring 42 x 38 mm, extending from L2 to L5 over 32 mm, causing a lifting of the skin plane without a defect. The mass is composed of fatty, radicular, and cerebrospinal fluid content, with a stretched appearance of the nerve roots of the cauda equina.

**Figure 1:** Spinal MRI in coronal T2, sagittal T1, T2, and fat-suppressed T1 with gadolinium injection sequences, revealing the presence of Bone defect of the posterior arches of the lumbosacral region extended from L4 to S1 with a large hernia of the dural sac containing cerebrospinal fluid, the nerve roots of the cauda equina and an infiltrating subcutaneous compressive fatty formation, Dorsolumbar scoliosis associated with vertebral malformations associating a rotational component, blind tiered hemivertebrae from D11 to L1 and fusion of the vertebral bodies of L1 and L2.

**Figure 2:** Spinal MRI in sagittal T1, T2, and fat-suppressed T1 with gadolinium injection sequences, revealing the presence of a herniated sac extending from a posterior arch defect over L2 to L5, causing a elevation of the skin plane. The mass consists of fatty, radicular, and cerebrospinal fluid (CSF) content, with a stretched appearance of the nerve roots of the cauda equina.

**DISCUSSION**

Lipomyelomeningocele is a rare birth defect of the spine. The most common clinical presentation is the presence of a lipomatous-looking mass located in the midline in the lumbosacral region [2]. However, this mass can be associated with other clinical signs of the skin such as tufts of hair, lumbosacral dimples, or cutaneous hemangiomas. Other no less frequent signs can also be objectified, in particular a motor deficit, sensory disorders, spasticity, urinary and urorectal...
sphincter disorders or orthopedic deformities [3]. The progressive nature of the onset of these symptoms is suggestive [4]. In our first case, it was a spinal deformity showing then the importance of thinking about this diagnosis in the face of any closed spinal deformity.

In the literature, lipomyelomeningocele appears to affect more females than males its prevalence is approximately 0.05 per 1,000. Lipomyelomeningoceles are half as common as lipomyeloceles [4]. Classically, 3 categories of lipomyelomeningocele are described according to the location: dorsal, transitional and caudal [2]. In our patients, it was a caudal type. However, cervical localization has been described by Heidari et al., [4].

The spinal ultrasound is considered the first-line imaging examination due to its readily available and non-invasive nature. It facilitates the detection of congenital spinal anomalies, especially occult dysraphisms.

MRI plays an essential role in the diagnosis and treatment of lipomyelomeningocele. MRI findings include expansion of the spinal canal and subarachnoid spaces, dorsal extension of the spinal cord and dura through spinal dysraphism. The stretched and deformed character of the marrow which faces the lipoma is a common sign. The T1 and T2 sequences are extremely essential, in particular for surgical management. They make it possible to determine the neurosurgical anatomy of the lesion and its relationship with neighboring structures. MRI also helps in detecting associated malformations of the spinal cord and the presence of syringomyelic cavities. The three-dimensional approach is very useful because it offers flexibility for performing multi-planar reconstructions, especially in patients with spinal deformity as is the case in our observation [1].

Since the natural progression of lipomyelomeningocele is the progression to neurological deficits and/or urological disorders, thus testifying to a fairly early diagnosis, and justifying the need to explore any occult dysraphism objectified from birth to avoid the development of complications that may be irreversible.

The goals of the surgery are to eliminate fatty masses in order to relieve the effect of attachment on the spine to preserve neuronal tissues [2].

**Conclusion**

Lipomyelomeningocele is a rare condition. It’s a lipoma that is tightly attached to the dorsal surface of a neural placode and extend dorsally through spina bifida to be continuous with subcutaneous fat. Imaging, in particular MRI, plays a major role in topographic diagnosis as well as in the analysis of relationships with neighboring structures in order to guide therapeutic management, which in general is surgical.

**References**