

Lipomeningocele in A 3-Year-Old Girl: A Case Report with initial Suspicion of Sacrococcygeal Teratoma

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ABSTRACT

BACKGROUND: Lipomeningocele is a complex form of closed spinal dysraphism that results from an error in early embryological development. It presents at birth as a lumbosacral mass and carries a high risk of progressive neurological, urological, and orthopedic deterioration due to the tethering of the spinal cord. While surgery is the primary treatment, the timing of intervention in asymptomatic patients is a key consideration.

CASE PRESENTATION: A 3-year-old girl presented with a large, skin-covered congenital mass in the lumbosacral region that had been present since birth. While motor and sensory functions were intact, neurological examination revealed bilateral Babinski reflexes, an early sign of neurological compromise. The diagnostic workup included an ultrasound and CT scan, which provided an initial differential diagnosis. A definitive diagnosis was established with a lumbosacral MRI, which confirmed a lipomeningocele with the conus medullaris tethered at the L5 level. The patient underwent surgical resection and defect closure. However, nine days postoperatively, she presented with wound dehiscence and a cerebrospinal fluid (CSF) leak, which necessitated a second surgical procedure for debridement and watertight dural repair.

CONCLUSION: This case highlights the essential role of MRI as the definitive imaging modality for diagnosing lipomeningocele and planning surgery. It reinforces the strong rationale for early surgical intervention, even in minimally symptomatic patients, to untether the spinal cord and prevent irreversible neurological decline. Finally, it serves as a critical reminder that this complex surgery carries significant risks, such as CSF leakage, which demand vigilant postoperative monitoring and prompt re-intervention.

INTRODUCTION

Introduction

Lipomeningocele is a complex form of closed spinal dysraphism, with a prevalence of approximately 0.6 per 10,000 live births.¹ It results from a critical error during early embryological development, specifically a premature disjunction of the epithelial ectoderm from the neural ectoderm.^{2,3}

This event allows mesenchymal cells, which later form fatty tissue, to fuse with the developing spinal cord.⁴ Both genetic and environmental factors are thought to contribute to its occurrence, and unlike other neural tube defects, its link to maternal folic acid deficiency is considered less clear.^{4,5}

The condition typically presents at birth as a subcutaneous fatty mass in the lumbosacral region, often accompanied by cutaneous markers such as skin dimples or an asymmetric gluteal cleft.^{6,7} This abnormal attachment inherently tethers the spinal cord, restricting its movement and leading to a high risk of progressive neurological, urological, and orthopedic deterioration as the child grows.^{1,7} This report presents the case of a 3-year-old girl with lipomeningocele, illustrating the diagnostic pathway, the rationale for early surgical intervention, and the management of a significant postoperative complication.

CASE PRESENTATION

We presented the case of a 3-year-old female who was initially referred to the neurosurgery department for evaluation of a large congenital mass on her lower back.

The patient's parents reported a lump on her lumbosacral region that had been present since birth and had progressively enlarged. Her history was negative for any discharge from the mass, bowel or bladder dysfunction, limb weakness, or seizures. She was born full-term at 40 weeks with a birth weight of 5,000 grams. On initial physical examination, the local examination revealed a large, 14x10x5 cm, well-defined, skin-covered mass in the lumbosacral area with associated skin dimpling. Neurologically, her motor and sensory functions were intact, but bilateral Babinski reflexes were present.

The diagnostic workup began with an ultrasound, which suggested a cystic sacrococcygeal



Figure 1. A large, 14x10x5 cm, well-defined, skin-covered mass in the lumbosacral area with associated skin dimpling.

teratoma. A subsequent CT scan, however, revealed a posterior spinal defect at L5-S1 consistent with a sacral meningocele. Definitive diagnosis was established by a lumbosacral MRI, which confirmed a lipomeningocele, characterized by a fatty mass extending through a vertebral defect. The MRI also demonstrated a low-lying conus medullaris at the L5 level, consistent with a tethered spinal cord.

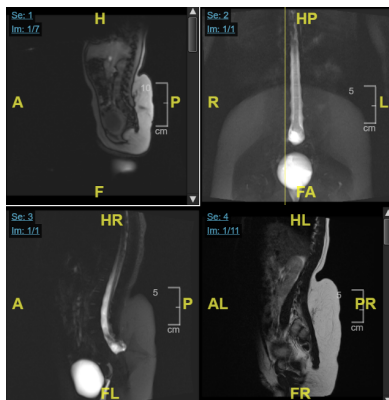


Figure 2. Lumbosacral MRI confirming a lipomeningocele with a low-lying conus medullaris at the L5 level, consistent with a tethered spinal cord.

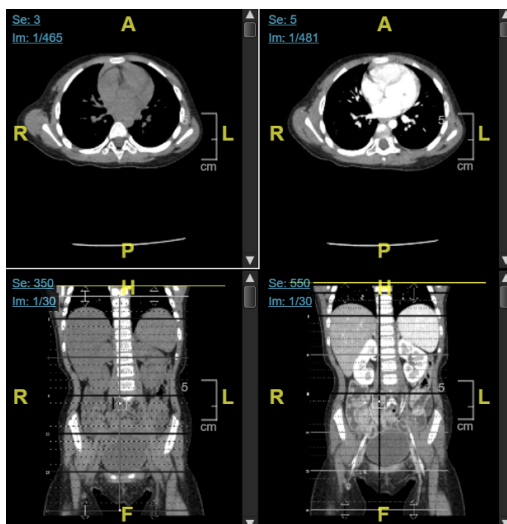


Figure 3. Abdominopelvic CT scan from the initial diagnostic workup. The imaging revealed a cystic mass communicating with the spinal canal through a posterior vertebral defect at L5-S1, leading to a preliminary diagnosis of sacral meningocele.

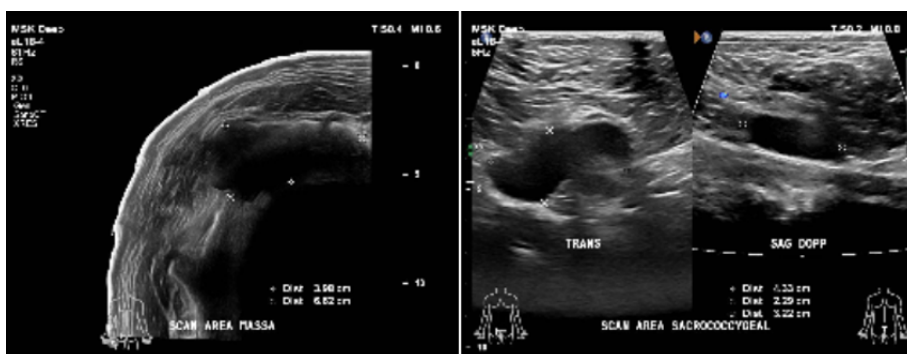


Figure 4. Initial ultrasound evaluation of the sacrococcygeal mass. The study revealed a cystic lesion, creating a differential diagnosis between a cystic sacrococcygeal teratoma and a meningocele, which prompted further advanced imaging.

Based on these findings, the patient was diagnosed with a lumbosacral lipomeningocele and underwent a planned surgical resection and defect closure. She was discharged five days later in good condition.

The resected tissue was sent for pathological analysis. Macroscopic examination revealed several fragments of amber-colored, skin-covered fatty tissue. Microscopic analysis confirmed the presence of mature adipose (fat) cells, disorganized fibrous tissue, blood vessels, along with bundles of smooth muscle and nerve fibers. No signs of malignancy were identified. The final pathological diagnosis was lumbosacral lipomeningocele.

Nine days postoperatively, the patient was brought to the emergency department with a new chief complaint of clear fluid leakage from the surgical wound. On this second admission, her physical examination revealed a 10x6 cm postoperative wound site with active leakage of a clear, serous fluid. Laboratory investigations were significant for leukocytosis (15,630/ μ L).

The patient was diagnosed with wound dehiscence with suspected cerebrospinal fluid (CSF) leakage. The immediate management plan included sending a swab of the wound discharge for culture and sensitivity, as well as testing the fluid for Beta-2 transferrin to confirm the presence of CSF. She was scheduled for urgent surgical intervention, involving wound debridement and watertight dural repair.

DISCUSSION

The management of lipomeningocele remains a significant challenge, requiring careful consideration of diagnosis, surgical timing, and postoperative risks. This case highlights several key aspects of this complex condition.

The Diagnostic Pathway and Role of Imaging

The patient's journey to a definitive diagnosis was structured. The presentation with a lumbosacral mass and skin dimple is a classic sign of a closed spinal dysraphism.^{3,7} The initial ultrasound created a differential diagnosis, which is common, as ultrasound is a valuable but limited screening tool.⁸ As supported by extensive literature, Magnetic Resonance Imaging (MRI) was the definitive imaging modality. It provided the crucial anatomical detail needed to confirm lipomeningocele, visualize the tethered cord, and plan the surgical strategy.^{8,9}

Surgical Management and Timing

Surgery is the mainstay of treatment for lipomeningocele, with the primary goals being the excision of the lipoma and untethering of the spinal cord, all while preserving neural function.^{5,7,10} The central debate revolves around when to operate on asymptomatic or minimally symptomatic infants. While a conservative approach has been considered, a strong consensus favors early intervention to prevent the onset of progressive, often irreversible deficits, particularly concerning bladder and bowel function.^{5,7,9}

Our patient, while largely asymptomatic, had bilateral Babinski reflexes, an early sign of neurological compromise. The decision to proceed with prophylactic surgery aligns with the principle that outcomes are significantly better when intervention occurs before major deficits are established.^{5,9} This is especially pertinent in regions where consistent long-term follow-up may be difficult, making early, definitive treatment a more secure strategy to prevent future decline.³

Postoperative Complications

While surgery aims to improve the natural history of lipomeningocele, it carries notable risks. The most common complications are wound infection and cerebrospinal fluid (CSF) leakage.^{5,7} In a regional study, Koirala et al. reported a wound infection rate of 23% in their series of 13 patients.⁵ Our patient developed a CSF leak nine days postoperatively, a serious complication that required urgent re-operation. This event underscores that the postoperative period is a critical phase of management. It highlights the absolute necessity for vigilant monitoring to promptly identify and manage such adverse events, which are an inherent risk of this complex spinal surgery.^{5,8}

CONCLUSION

Lipomeningocele is a complex congenital anomaly that requires a comprehensive approach from diagnosis through long-term follow-up. This case effectively demonstrates the essential role of a multi-modality imaging pathway, culminating in MRI for definitive diagnosis and surgical planning. Furthermore, it reinforces the rationale for early surgical intervention aimed at untethering the spinal cord to prevent irreversible neurological decline. While the long-term outcomes of surgery are generally favorable, this case also serves as a critical reminder that the procedure carries significant risks. Postoperative complications like CSF leakage are not uncommon and demand vigilant care and the readiness for prompt re-intervention.

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