

**Lateral meningocele syndrome in a Nigerian child: a case report and literature review**

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**ABSTRACT**

Lateral meningocele syndrome is a very rare neurosurgical disorder characterized by the presence of multiple lateral thoracolumbar spinal meningoceles. Very few cases have been reported in literature and as such there are no standard management protocols and treatment is still fraught with several controversies. We present a 2-year-old girl with a 16-month history of progressive abdominal distention associated with occasional abdominal pain. Thoracoabdominal magnetic resonance imaging showed a huge left-sided cystic intra-abdominal mass lesion with a fistulous connection to the spinal subarachnoid space at the level of the third lumbar (L3) vertebra and other smaller cystic thoracolumbar lateral meningoceles. She had L3, L4 left hemi-laminectomy, decompression of the cyst and obliteration duroplasty to close the fistulous connection. Post-operative period was however complicated by cyst re-accumulation, pseudomeningocele and meningitis. Follow-up neuroimaging revealed hydrocephalus necessitating ventriculoperitoneal shunt insertion. This case highlights a very rare congenital anomaly and the attendant diagnostic and management challenges.

**Keywords:** Lateral meningocele syndrome, obliteration duroplasty, pseudomeningocele, cerebrospinal fluid

**INTRODUCTION**

Lateral meningocele syndrome (LMS) is a rare neurosurgical disorder.<sup>1</sup> It has been described as a genetic connective tissue disorder with morphological changes similar to those seen in other connective tissue disorders.<sup>1,2</sup> Its morphological hallmarks are multiple bilateral, large lateral meningoceles herniating through the spinal foramina.<sup>3</sup> These lateral meningoceles can occur primarily as seen in Lehman syndrome, also known as LMS or as a component of other connective tissue disorders like Marfan syndrome, Nevo syndrome, Hajdu-Cheney syndrome as well as Neurofibromatosis type 1 (NF1).<sup>3</sup> These meningoceles may remain small and asymptomatic but can also become large and symptomatic.<sup>4</sup> In such cases, the patient may present with axial or radicular back pain, discomfort and motor weakness due to direct compression of the spinal nerve root exiting the foramen; there may also be abdominal distention and respiratory embarrassment from mass effect on retroperitoneal and intrathoracic structures.<sup>5-7</sup> Lateral meningoceles often distort the cerebrospinal fluid (CSF) flow dynamics and tend to co-exist with concurrent Chiari malformation thus creating unique management challenges.<sup>3,8</sup>

We present a 2-year-old female toddler with multiple lateral meningoceles with a symptomatic giant lateral lumbar meningocele who developed hydrocephalus necessitating CSF diversion following surgical repair of the lumbar intra-abdominal lesion.

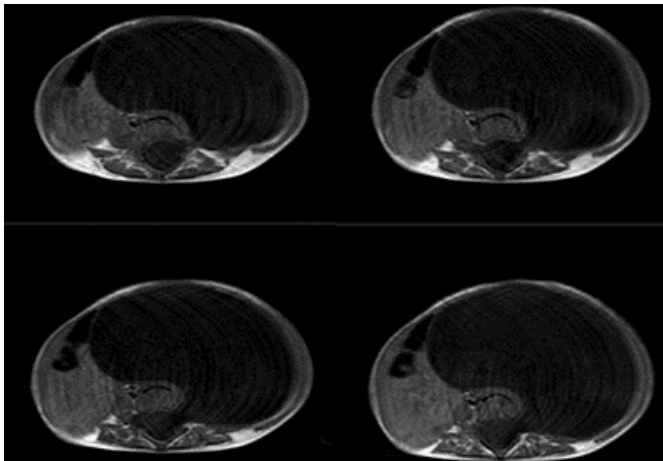
**CASE**

A 2-year-old female toddler presented to us with a history of progressive abdominal distention since the 8<sup>th</sup> month of life. There was occasional history of abdominal pain. However, there was no vomiting, early satiety, constipation or lower urinary tract symptoms. There was no history of chest pain, cough, breathlessness or weight loss. She had no limb weakness or sphincter dysfunction. She was a product of an uneventful term gestation delivered via spontaneous vaginal delivery. Physical examination revealed a fully conscious child with dysmorphic facies (arched eyebrows, flattened midface, and angulated eyes and ears). Neurological examination was essentially normal. The abdomen was grossly distended without any palpably enlarged organs.

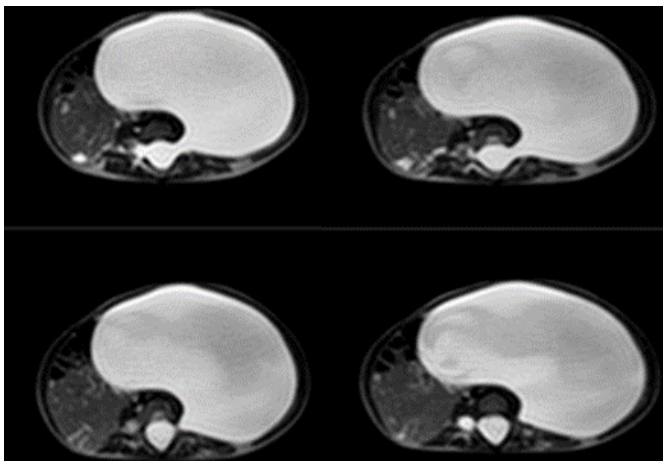


Percussion note was dull and bowel sound was normoactive. She had normal female external genitalia and no stigmata of a neurocutaneous syndrome. Her laboratory tests revealed no abnormal findings.

Thoracoabdominal magnetic resonance imaging (MRI) showed multilevel bilateral thoraco-lumbar lateral paraspinous outpouchings one of which was huge and dominant—a left-sided cystic lesion communicating with the spinal subarachnoid space at the L3 level which extended into the retroperitoneum with displacement of bowel loops superiorly and inferiorly. The cyst was hypointense on T1-weighted and hyperintense on T2-weighted images (Figures 1-4).

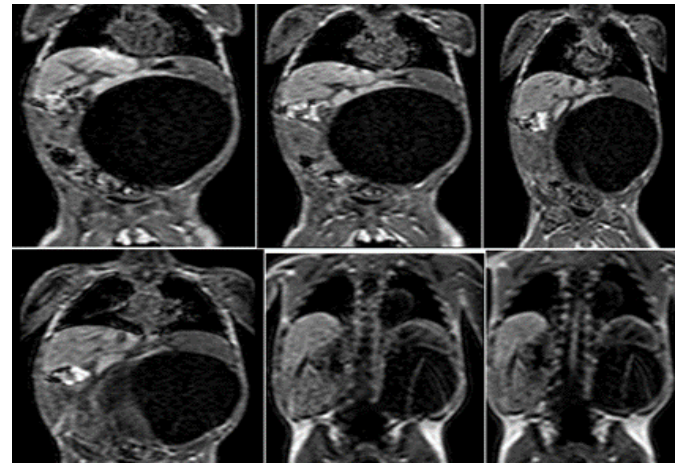


**Figure 1.** Axial T1-weighted images showing a left-sided hypointense cystic lesion communicating with the spinal subarachnoid space

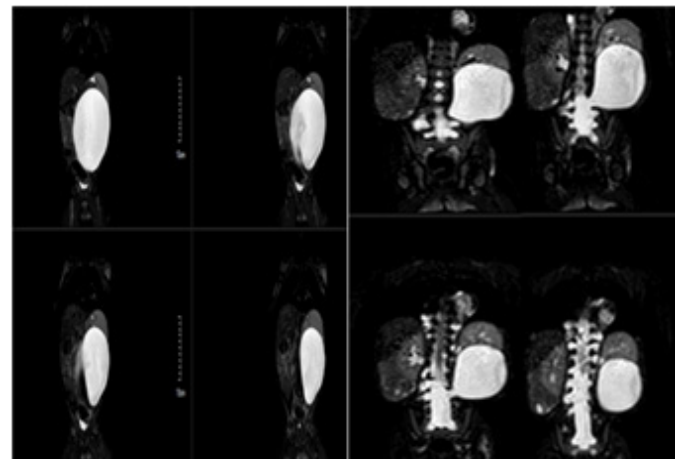


**Figure 2.** Axial T2-weighted images showing a left-sided hyperintense cystic lesion communicating with the spinal subarachnoid space

The patient had no clinical features of an intracranial pathology or elevated intracranial pressure; hence a cranial imaging was not done at this time. She had L3, L4 left hemilaminectomy and durotomy with intraoperative findings of egress of clear CSF under marked pressure from the dominant meningocele via a 4cm wide fistula between the cyst and the spinal subarachnoid space. The fistula was located at the lateral aspect of the L3 vertebra. The meningocele was decompressed by manual pressure on the abdomen till there was no longer egress of CSF via the fistula. About 1000mls of CSF was drained. An obliterative duroplasty was done to close the CSF fistula followed by a water-tight duroplasty. Multiple blind ending outpouchings of the dura were also noted intraoperatively. Immediate



**Figure 3.** Coronal T1-weighted images showing the same lesion communicating with the spinal subarachnoid space at the L3 level



**Figure 4.** Coronal T2-weighted images showing the same lesion communicating with the spinal subarachnoid space at the L3 level

post-operative period was uneventful, bowel sound was normoactive on the first post-operative day and feeding was commenced. One week post-operatively, she had a re-accumulation of the cyst with progressive abdominal expansion necessitating wound exploration, cyst drainage and a reinforcement of the obliteration duroplasty. Following the re-do surgery, the abdominal swelling never recurred. She subsequently developed pseudomeningocele, which resolved with aseptic aspiration and firm dressing. After resolution of the pseudomeningocele, she developed meningitic features i.e. fever, nuchal rigidity and seizures. The diagnosis of bacterial meningitis was confirmed via CSF culture and she was managed with appropriate antibiotics by the Pediatricians. She subsequently developed increased seizure frequency with subsequent status epilepticus in spite of combination anti-seizure medications necessitating cranial computed tomographic (CT) scan and admission into the Intensive Care Unit. The cranial CT scan revealed communicating hydrocephalus. She had CSF diversion via an initial external ventricular drain and a definitive ventriculo-peritoneal shunt insertion after serial negative CSF cultures. Fever and seizures however persisted, her clinical condition continued to deteriorate until she died about 8 weeks after the first surgery. Parents declined autopsy.

## DISCUSSION

LMS is characterized by lateral meningoceles herniating through the intervertebral foramina and associated scalloping

of the posterior vertebral bodies.<sup>5,7</sup> The meningoceles are mostly dependent, occurring in the lower thoracic and lumbar spine, unlike in NF1 where they mostly occur in the thoracic spine.<sup>3,9</sup> Lateral meningoceles can occur in isolation or as a component of other connective tissue disorders or syndromes. Such syndromes include Loeys-Dietz syndrome, Marfan syndrome, Ehler Danlos syndrome, Hajdu-Cheney syndrome, Nevo syndrome and Lehman syndrome.<sup>9,10</sup> Our patient had some dysmorphic facial features which suggested that the lateral meningocele was likely syndromic. Confirmation of syndromic association is usually done via genetic tests which are not readily available in our setting. For example heterozygous, de novo 2-nucleotide deletion in exon 33 on the NOTCH3 gene is diagnostic of LMS.<sup>11,12</sup> While many lateral meningoceles may be asymptomatic, some may become large enough and cause symptoms via mass effect on nerve roots, the spinal cord or adjoining structures manifesting as abdominal distention and/or respiratory distress.<sup>3,13,14</sup> Our patient had progressive abdominal distention and abdominal pain. LMS though congenital may become symptomatic for the first time in adult life.<sup>6,8</sup> The likely diagnosis of LMS is usually suggested following neuroimaging like thoracolumbar MRI as seen in our patient. Some authors recommend imaging the entire neuroaxis because of other possible co-existent pathologies which may not be symptomatic at initial presentation but may impact on management.<sup>4,10</sup> Such pathologies include hydrocephalus, Chiari 1 malformation, kyphoscoliosis and syringomyelia among others.<sup>4,10</sup> Our patient had recurrent pseudomeningocele following surgical intervention and subsequently developed features of raised intracranial pressure. Cranial CT scan done at this time showed active hydrocephalus warranting CSF diversion. Even though there was no preoperative cranial imaging, we think the hydrocephalus may have been present but latent prior to surgery but subsequently became active after surgical intervention. It was thought that ligating a huge lateral meningocele like the one in this patient may disrupt the CSF dynamics causing active hydrocephalus.<sup>3</sup>

What constitutes optimal neurosurgical management in these patients is still poorly defined largely due to the rarity and variable clinical presentation.<sup>9</sup> The various interventions that have been reported in literature include laminectomy, decompression and ligation/obliteration as seen in this patient, ventriculoperitoneal shunt insertion, lumboperitoneal shunt and suboccipital craniectomy for foramen magnum decompression in co-existent chiari 1 malformation.<sup>2,3,9</sup> Patients with multilevel bilateral symptomatic meningoceles which may preclude laminectomy and ligation have been shown to improve with CSF diversion alone.<sup>2</sup>

Complications associated with treatment include pseudomeningocele as seen in our patient and spine destabilization needing instrumented spine fusions, due to the presence of bony changes of the spine such as scoliosis and scalloped vertebrae.<sup>3,9</sup>

## CONCLUSION

This case highlights a neurosurgical rarity-LMS and the attendant management challenges due to the absence of standard treatment protocols. It is hoped that that more clarity is gained with each reported case.

## ETHICAL DECLARATIONS

### Informed Consent

Written informed consent was obtained from the parents of the patient included in this report. Signed consent forms are retained by the authors and are available upon request.

### Peer Review Process

This report underwent external peer review.

### Conflict of Interest

The authors declare no conflicts of interest.

### Financial Disclosure

This case report did not receive any financial support.

### Author Contributions

Concept: ASY, OOA; Design: ASY, OOA; Supervision: ASY; Literature Review: ASY, MMI, OOA; Manuscript Preparation: All authors; Critical Review: All Authors.

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