

PAKISTAN JOURNAL OF NEUROLOGICAL SURGERY (QUARTERLY) – OFFICIAL JOURNAL OF PAKISTAN SOCIETY OF NEUROSURGEONS



Case Report

Anterior Lumbosacral Myelomeningocele Presenting as an Abdominal Mass

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ABSTRACT

Myelomeningocele is a complex pathology associated with variable clinical presentations and associated anomalies. Myelomeningocele presenting as an abdominal mass is extremely rare. This case report presents such a complex case of anterior lumbosacral myelomeningocele with clinical presentation of abdominal mass along with its surgical management. A 5-month-old boy was referred to the neurosurgery department, presenting with a congenital lump on his lower back along with abdominal mass, constipation, and urogenital dysfunction. An anterior Approach exploratory laparotomy was performed by a left lower abdomen oblique incision to excise the myelomeningocele. This case report illustrates that anterior lumbosacral myelomeningocele can present, in rare cases as abdominal mass along with urogenital dysfunctions and constipation and is managed with a collaborative effort from a multidisciplinary approach including urologists, neurosurgeons, and general surgeons.

Keywords: Anterior Lumbosacral Myelomeningocele, Abdominal Mass, Lumbosacral Myelomeningocele, Spina Bifida.

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Date of Submission: 12-06-2023
Date of Revision: 31-08-2023
Date of Acceptance: 25-09-2023
Date of Online Publishing: 30-09-2023

Date of Print: 30-09-2023

DOI: 10.36552/pjns.v27i3.868

INTRODUCTION

Myelomeningocele represents the most severe form of spina bifida and is characterized by motor and sensory defects, genitourinary dysfunctions, and hydrocephalus. The symptoms manifested due to protrusion of the spinal cord and meninges through a defect in the vertebral arch. The incidence of myelomeningocele is reported to be between 0.2 and 2 per 1000 live births.¹ Certain factors contribute to development of myelomeningocele including genetic predisposition, low socioeconomic status, maternal diseases, and deficiency of folic acid. The most common region of the spine affected is the lumbosacral area followed by the thoracic spine.² Myelomeningocele is categorized as a defect. associated complex with concurrent clinical anomalies and varied clinical presentations, the most common of which are Arnold Chiari malformation II and hydrocephalus. screening with alpha-fetoprotein, ultrasonography, and a visible bulge on the back is used to diagnose the condition.³ Surgical repairs are the treatment of choice to repair or close the neural defect while additional surgeries are also required to manage the concurrent anomalies.4

This case report presents the case of a 5 5-month-old child diagnosed with anterior lumbosacral myelomeningocele presenting with an abdominal mass. The purpose of this study was to describe the variable clinical presentation associated with the most severe form of spina bifida along with its surgical management.

CASE PRESENTATION

History of Presenting Condition

We report a rare case of a 5-month-old child who presented to the neurosurgery department with a congenital lump on the lower back along with an anterior abdominal mass. The parents had noticed the lump since birth which was soft and increasing in size with no urinary incontinence or retention. The child had constipation and abdominal fullness. There was no history of birth trauma or any surgical intervention.

Examination Findings

On examining the patient, a 2.5×2.5 cm firm circular lump was observed on the lumbosacral area which was bluish in color, non-tender, non-blanching, and non-fluctuant. Transillumination revealed no cystic mass posteriorly. On abdominal examination, it was found that the abdomen was distended, soft, non-tender, and

bowel sounds were audible with the frequency of 12 per minute having normal pitch. A digital examination revealed no anorectal malformation along with normal anal tone. Chest examination showed normal heart and lung sounds along with normal chest x-ray with no abnormality. The echocardiogram revealed no congenital aberrant or malformations. Examination of the back and spine revealed no bony defects posteriorly and no neurological deficit was detected.



Figure 1: Sacral lump on observation (Image included with patient consent).

The hematological profile of Hemoglobin, Total leukocyte count, and platelet level was also within normal limits.

Magnetic resonance imaging of the abdomen and back revealed a presacral cystic mass communicating with the spinal dural sac through a neck traversing the sacral bony defect, extending to L1 occupying the majority of the lower abdomen, hyper dense & compressing the surrounding bowel and bladder. Scimitar sacrum was present while the Currarino triad were absent.

Diagnosis and Management

As the MRI findings correlated with the subjective and objective examination, the diagnosis of

Pre-Operational MRI

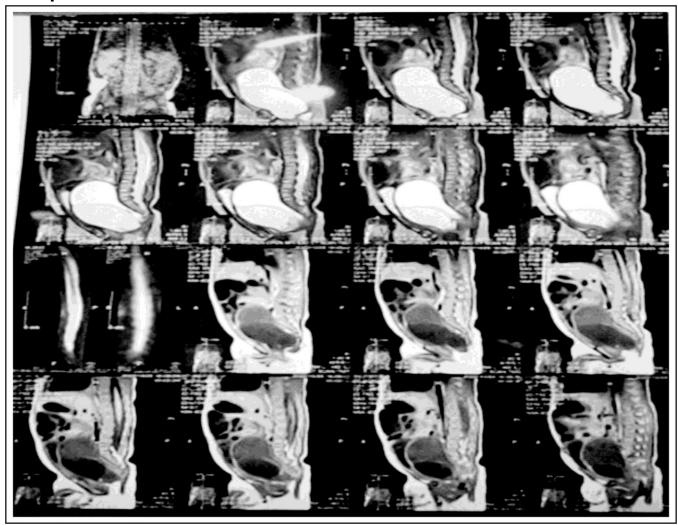


Figure 2 (a): Pre-operative Sagittal lumbar spine MRI demonstrating the location of myelomeningocele (Images included with patient consent).

anterior lumbosacral myelomeningocele was made. Parents of the child were counseled about the condition and treatment options. By looking at the extent and chronicity of the condition, it was decided to perform the Anterior Approach exploratory laparotomy.

Intervention

The patient was given general anesthesia and catheterized with a silicone catheter. An Anterior Approach exploratory laparotomy was performed by a left lower abdomen oblique incision to excise the myelomeningocele. A cystic lesion measuring

about 6×5 cm was seen emerging from the anterior part of the sacrum in the pelvis. The sigmoid colon was pushed to the right while the urinary bladder was pushed to the left side. The sac was identified, mobilized, and aspirated for cerebrospinal fluid. The sac was opened at the apex and then dissected till the base, purse string suture was used and the sac was ligated at the neck. The closed dural defect was sequentially checked for leakage of CSF and hemostasis was established. An intra-abdominal drain was placed and a wound was closed in 3 layers. Sac was sent for a histopathological examination.



Figure 2 (b): Pre-operative axial-lumbar spine MRI demonstrating the location of myelomeningocele (Images included with patient consent).

DISCUSSION

Myelomeningocele is a frequently reported condition presenting in pediatric neurosurgery often characterized by significant and disabilities such as sexual and sphincteric dysfunctions and sometimes paraplegia if the neural structures of the spinal cord are compromised.⁵ Myelomeningocele, occurring early in gestation is the most severe form of spina bifida characterized by complex congenital nonclosure of neural tube, resulting in exposed nervous tissue on the patient's back.⁶ Young children mostly present with urinary symptoms or constipation, whereas in adults it can present as

lower back and pelvic pain, constipation, difficulties in defecation, dysmenorrhea and dyspareunia, and urinary incontinence, retention, or urgency.⁷⁻⁸

While the lumbosacral area is most commonly affected by myelomeningocele as is consistent with our study, the location of the spine affected by myelomeningocele may vary. In a study conducted in Hungarian in which 352 cases of myelomeningocele were observed, the areas affected were follows: sacral (34.5%),as lumbosacral (22.3%),lumbar (16.8%),thoracolumbar (17.4%),thoracic (4.2%),cervicothoracic (0.9%) and cervical (1.8%) showing

Post-Operative MRI

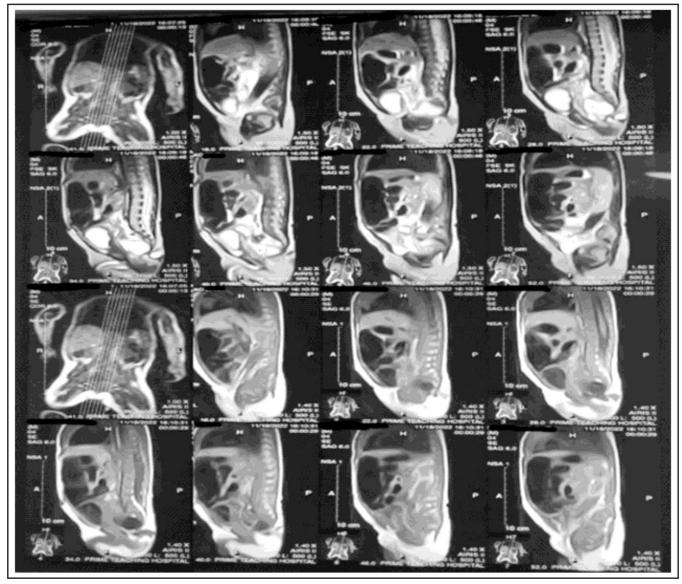


Figure 3: Post-operative MRI showing excision of myelomeningocele (images included with patient consent).

the heterogenicity of the areas affected by myelomeningocele.9

A case report conducted in Michigan reported the case of a Caucasian female diagnosed with anterolateral lumbar lipomyelomeningocele in which the patient presented with expanding abdominal mass along with other defects including urogenital system. The findings are consistent with our case report indicating that myelomeningocele in rare cases can present as

abdominal mass along with other clinical manifestations.¹⁰

The optimum time for surgical repair of myelomeningocele is within the first 48 hours after birth to preserve neurological structures integrity and further complications. Delayed surgical procedures impose negative effects on patients' outcomes and disease prognosis.¹¹ Management of myelomeningocele requires a holistic approach and lifelong input from different

health disciplines including general surgeon, urologist, orthopedics, physical and social therapy beside neurosurgery. The most important step in the surgical treatment is to repair and reverse the complications and abnormal anatomy associated with the failed closure of the neural tube.12 Different surgical approaches are used for the repair and management of myelomeningocele such as primary repair closure and V-Y plasty.¹³ Since a large presacral mass was present in our anterior Approach exploratory case, an laparotomy was performed by a left lower oblique incision abdomen to excise myelomeningocele.

CONCLUSION

Myelomeningocele is a complex pathology associated with variable clinical presentations and different surgical options. This case report illustrates that anterior lumbosacral myelomeningocele can present, in rare cases as abdominal mass along with urogenital dysfunctions and constipation and is managed with a collaborative effort from a multidisciplinary approach including urologists, neurosurgeons, and general surgeons.

REFERENCES

- McCarthy DJ, Sheinberg DL, Luther E, McCrea HJJNf. Myelomeningocele-associated hydrocephalus: nationwide analysis and systematic review. 2019;47(4):E5.
- 2. Kitov B, Kehayov IJFm. Myelomeningocele with Associated Anomalies–Case Report and Literature Review, 2019;61(3):462-6.

- 3. Ntimbani J, Kelly A, Lekgwara PJIN. Myelomeningocele-A literature review, 2020:19:100502.
- Paslaru FG, Panaitescu AM, Iancu G, Veduta A, Gica N, Paslaru AC, et al. Myelomeningocele Surgery over the 10 Years Following the MOMS Trial: A Systematic Review of Outcomes in Prenatal versus Postnatal Surgical Repair, 2021;57(7):707.
- Anegbe AO, Shokunbi MT, Oyemolade TA, Badejo OA. Intracranial infection in patients with myelomeningocele: profile and risk factors. Child's Nervous System, 2019;35(11):2205-10.
- 6. Jowdy PK, Perry RJ, Reynolds RMJJoNP. Use of an abdominal reapproximation anchor system in the closure of large, open myelomeningoceles, 2020;25(4):402-6.
- 7. Bedi NK, Chadha R, Bagga D, Anand R, Mohta A. Anterior sacral meningocele: an uncommon cause of constipation in early childhood. Indian pediatrics, 1992;29(9):1157-60.
- 8. Ashley WW, Jr., Wright NM. Resection of a giant anterior sacral meningocele via an anterior approach: case report and review of literature. Surgical neurology, 2006;66(1):89-93; Discussion.
- Ntimbani J, Kelly A, Lekgwara P Myelomeningocele - A literature review Interdisciplinary Neurosurgery, 2020;19:100502.
- 10. Barami K, Pereira J, Canady Al. Anterolateral lumbar lipomyelomeningocele: case report and review of the literature. Neurosurgery, 1997;41(6):1421-3; Discussion 3-4.
- 11. Diao Y, Rong H, Zhu T. Adult lumbosacral spina bifida with giant myelomeningocele: a case report. Acta Neurologica Belgica. 2022.
- 12. Gabrielsson H, Riedel D, Bendt M. Living with Spina Bifida myelomeningocele as adults. 2022.
- Lobo GJ, Nayak M. V-Y Plasty or Primary Repair Closure of Myelomeningocele: Our Experience. Journal of Pediatric Neurosciences, 2018;13(4):398-403.

Additional Information

Disclosures: Authors report no conflict of interest.

Ethical Review Board Approval: The study conformed to the ethical review board requirements.

Human Subjects: Consent was obtained by all patients/participants in this study.

Conflicts of Interest:

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous

three years with any organizations that might have an interest in the submitted work.

Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHORS CONTRIBUTIONS

Sr. No.	Author's Full Name	Intellectual Contribution to Paper in Terms of
1	Mumtaz Ali & Abdul Haseeb Sahibzada	Study Design, Methodology, and Paper Writing
2	Akram Ullah & Mumtaz Ali	Data Calculation and Data Analysis
3	Hanif Ur Rahman	Interpretation of Results
4	Abdul Haseeb Sahibzada	Statistical Analysis
5	Summayah Saqib & Amjad Ali	Literature Review
6	Mumtaz Ali	Literature Review and Quality Insurer