Giant Thoracolumbosacral Myelomeningocele: A Case Report

Abhijit Acharya¹⁰ Souvagya Panigrahi¹ Rama Chandra Deo¹ Satya Bhusan Senapati¹ Ashok Kumar Mahapatra¹

¹ Department of Neurosurgery, IMS and SUM Hospital, SOA University Kalinga Nagar, Bhubaneswar, Odisha, India

Indian | Neurosurg

Abstract

Address for correspondence Abhijit Acharya, MCh, Department of Neurosurgery, IMS and SUM Hospital, Bhubaneswar, Odisha, India (e-mail: abhijitkirtika@gmail.com).

Neural tube defects are a broad spectrum of congenital developmental anomalies that include cranial defects and open and closed spinal dysraphism. Myelomeningocele is a common form of spinal dysraphism most commonly seen in the lumbosacral region. Giant myelomeningoceles are rare occurrences. We hereby present the case of a 7month-old infant with a history of large swelling over the thoracic lumbar region (circumference of 42 cm and diameter of 12.5 cm) that was progressively increasing in size with occasional cerebrospinal fluid (CSF) leakage from the swelling. There was associated progressive increasing size of the head size circumference (51 cm) as well for the past 6 months. There was no movement of both lower limbs since birth without any sensation up to the groin. On evaluation, the infant was found to have gross hydrocephalous. The Evans index was 0.4 with Chiari 2 malformation and a giant thoracolumbosacral myelomeningocele. The patient underwent a low-pressure ventriculoperitoneal (VP) shunt on the right side, followed by the repair of the giant myelomeningocele. The post-op period was uneventful with no CSF leakage or skin necrosis. The infant was discharged on day 5. Giant myelomeningoceles are guite a challenge for neurosurgeons due to the high risk of skin edge necrosis, CSF leaks, myelomeningocele meningoencephalitis, etc. Here we managed our case with utmost intraoperative ► neurosurgery surgical skills and postoperative management due to which our patient was discharged spinal dysraphism in a satisfactory condition without any complication. This case is supposed to be largest tethered cord myelomeningocele in the world literature to date.

Introduction

Keywords

The commonest area of development of spinal dysraphism is the lumbosacral region followed by the cervical and thoracic regions. Large myelomeningoceles involving the entire thoracic, lumbar, and sacral regions are quite rare.^{1,2} The myelomeningoceles were found to have multi-organ involvement like hydrocephalous, Chiari malformation, myelopathy, bladder and bowel sphincter involvement, and renal, respiratory, and cardiac complications. Other clinical

> DOI https://doi.org/ 10.1055/s-0044-1780520. ISSN 2277-954X.

features experienced in childhood include pain, hypertonia, vertebral anomalies, tethered cord, and psychological and cognitive complications.³

Case Report

A 7-month-old infant presented with a gradually progressive large swelling over the lumbosacral region extending over the entire thoracolumbosacral region since birth. It was also associated with a gradually progressive head circumference

This is an open access article published by Thieme under the terms of the Creative Commons Attribution License, permitting unrestricted use, distribution, and reproduction so long as the original work is properly cited. (https://creativecommons.org/licenses/by/4.0/) Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

^{© 2024.} The Author(s).

for the past 6 months. The infant was the second child from a nonconsanguineous marriage delivered by cesarean section at an institution. The parents' elder child was 7 years old and healthy. The infant cried immediately after birth and passed urine and stools normally. The infant was moving the upper limbs normally; however, the lower limbs were not moving at all and were lying flaccid. Clinical examination revealed both lower limb power to be 0/5 (as per the Medical Research Council [MRC] grade) with complete sensory loss of all modalities up to the groin. The parents did seek neurosurgical consultation and were advised for surgery soon after the birth of the baby, but they deferred it due to lack of money and the life risk involved. There was an occasional history of leakage of fluid from the swelling. The swelling was approximately $20 \text{ cm} \times 15 \text{ cm} \times 12 \text{ cm}$ in size, with a circumference of 42 cm, lobulated, with a broad base extending from the thoracic to the sacral area, and covered with thick desquamated epithelium (**Fig. 1**). The infant was voiding urine spontaneously and intermittently with occasional dribbling with a history of urinary tract infection. The circumference was of the infant's head was



Fig. 1 A giant thoracolumbosacral myelomeningocele.



Fig. 2 The child's large head circumference with frontal bossing with open anterior fontanelle.

51 cm (~20 inches) with frontal bossing with the anterior fontanelle open (> Fig. 2). Computed tomography (CT) of the spine was suggestive of a defect in the D10 to S1 vertebrae in the midline posterior aspect with a large cyst 15×12 cm posterior to the defect suggestive of spina bifida with myelomeningocele (**Fig. 3**). Magnetic resonance imaging (MRI) of the brain and the spine suggested a large swelling with few septations with nerve roots. Most parts of the swelling were T2 hyperintense on MRI of the spine suggestive of cerebrospinal fluid (CSF) collection and a part of the thoracic cord entering into the neck swelling (Fig. 4). MRI of the brain was suggestive of Chiari II malformation with gross hydrocephalous (Fig. 5). After a preoperative evaluation, the patient was planned for definitive surgery. А right-sided low-pressure ventriculoperitoneal shunt (Chhabra shunt) was placed. After the shunt placement, the infant was made to lie prone. A longitudinal incision was made involving the large swelling saving as much healthy skin as possible. The dissection was done all around the myelomeningocele sac at its junction with normal skin. The anatomy was distorted and the dural covering was friable and deficient at most of the part of the swelling. The neural placode was identified along with its ventral roots. The neural placode was seen at the base of the swelling with the normal cord proximal to it. The pial layer of two sides was sutured in the midline. Watertight dural closure was done to its maximum feasibility. A multilayered closure technique was adopted by closing the muscle, thoracolumbar fascia subcutaneous tissue, and skin (release incisions were given to approximate the layers without tension; **Fig. 6**). The postoperative course was nonsignificant (without any infection). The neurological status in the postoperative period was the



Fig. 3 Computed tomography (CT) of the spine suggestive of giant meningomyelocele.



Fig. 4 Magnetic resonance imaging (MRI) of the spine shows T2 hyperintensity of the sac suggestive of cerebrospinal fluid (CSF) collection and a part of the thoracic cord entering into the neck swelling.



Fig. 5 Magnetic resonance imaging (MRI) of the brain was suggestive of Chiari II malformation with gross hydrocephalous.



Fig. 6 Postoperative image of the surgical site.

same as before. The wound at the surgical site healed satisfactorily after 3 months (**Fig. 7**).

Technical Nuances

Primary closure of the skin was done, so that as much skin as possible could be preserved during surgery. The dura was opened at the normal area above, the nerve root was identified at the placode, and the filum was then identified and cut to prevent re-tethering.

Conclusion

This case is supposed to be the largest myelomeningocele reported in the world literature. Very few cases of this size have been reported in the medical literature. These patients are neglected and not operated on early. Such large-size



Fig. 7 Follow-up image of the surgical site (after 3 months).

myelomeningoceles will have a high risk of anesthesia complications, high blood loss because due to the young age, long duration of surgery of around 4 to 5 hours (given the large size), and hypothermia.

The senior author of the study has vast experience in the surgical management of all varieties of neural tube defects with satisfactory outcomes in most patients.⁴

Giant myelomeningoceles are a challenge for neurosurgeons due to the high risk of skin necrosis, CSF leak, meningoencephalitis, etc. In our patient, the circumference of the swelling was almost the size of the normal head of a baby. Utmost surgical skill with postoperative management was the key to the successful management of the case reported in this study.

Authors' Contribution

All the authors contributed to the study's conception and design. A.A. contributed to the conceptualization of the study and writing of the first draft. A.K.M. was responsible for preparation of the figures. S.P. contributed to the editing and writing of the manuscript. R.C.D. contributed to the editing of the manuscript. S.B.S. contributed to the editing and the second draft of the manuscript.

Conflict of Interest None declared.

Acknowledgment

We thank the SOA University (IMS & SUM Hospital), Bhubaneswar, Odisha, for allowing us to conduct this study.

References

- 1 Agrawal A, Sampley S. Spinal dysraphism: a challenge continued to be faced by neurosurgeons in developing countries. Asian J Neurosurg 2014;9(02):68–71
- 2 Patnaik A, Mahapatra AK. A rare case of giant multiseptated thoracic myelomeningocele with segmental placode. Surg Neurol Int 2015;6:170
- 3 Bakketun T, Gilhus NE, Rekand T. Myelomeningocele: need for long-time complex follow-up-an observational study. Scoliosis Spinal Disord 2019;14:3
- 4 Mahapatra AK. Spinal dysraphism controversies: AIIMS experiences and contribution. Indian J Neurosurg 2012