Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Report

Isolated Anterior Thoracic Myelomeningocele: A Rare Case Report

Vignesh Gadupudi[1], Pravalya Chaparala[2], Abhinay Sai Vidavaluru[3]

[1]Senior Resident, Department of Radio-diagnosis, A. C. S Medical College and Hospital, Chennai, Tamil Nadu, India, [2]Junior Resident, Department of Radio-diagnosis, Alluri Sitarama Raju Academy of Medical Sciences, Andhra Pradesh, India, [3]Junior Resident, Department of Radio-diagnosis, A. C. S Medical College and Hospital, Chennai, Tamil Nadu, India.

Abstract

Context: Myelomeningocele (MMC) is a type of spinal dysraphism that often affects the lumbosacral area with a rare thoracic form of presentation. Spinal abnormalities brought on by spinal dysraphism present a difficult issue. In the literature that is currently available, only few cases have been described. Often found posteriorly in the lumbosacral region, a spinal meningocele is a herniation of the meninges through a foramen or a defect in the vertebral column. The majority of documented thoracic meningoceles are not only anterior but also lateral or anterolateral in position. In this case report, a genuine anterior thoracic meningocele without extensive mesenchymal dysplasia is described. These rarely present alone, but commonly as a symptom of a widespread mesenchymal dysplasia such Marfan syndrome or neurofibromatosis type 1.

Case Report: A 2-year-old child presented with swelling in the upper thorax and with gradual onset upper limb weakness. Scoliosis was present. Magnetic resonance imaging (MRI) of spine showed anterior herniation of spinal cord and meninges through the bony defect in thoracic region. MRI imaging with its excellent soft tissue contrast helps in accurately mapping the anatomy and enables surgical planning.

Conclusion: Spinal cord lesions can be diagnosed on MRI. MRI imaging with its excellent soft-tissue contrast helps in accurately mapping the anatomy and enables surgical planning. The case report's goal is to raise awareness of the MMC's uncommon thoracic spine presentation and the possibility of scoliosis association with the lesion.

Key words: Butterfly vertebrae, Magnetic resonance imaging, Myelomeningocele, Thoracic

INTRODUCTION

Myelomeningocele (MMC) is a type of spinal dysraphism that often affects the lumbosacral area with a rare thoracic form of presentation. Spinal abnormalities brought on by spinal dysraphism present a difficult issue. In the literature that is currently available, only few cases have been described. Often found posteriorly in the lumbosacral region, a spinal meningocele is a herniation of the meninges through a foramen or a defect in the vertebral column. The majority of documented thoracic meningoceles are not only anterior but also lateral or anterolateral in position. In this case report, an anterior thoracic meningocele without extensive mesenchymal dysplasia is described. These rarely present alone, but commonly as a symptom of a widespread mesenchymal dysplasia such Marfan syndrome or neurofibromatosis type 1 (NF-1).

In this case, a 2-year-old boy had edema in his upper thorax and gradually developed upper limb paralysis. The case report's goal is to raise awareness of the MMC's uncommon thoracic spine presentation and the possibility of scoliosis association with the lesion.

CASE REPORT

A 2-year-old boy complained of palpitation and shortness of breath, which were exacerbated by physical activity. He had diminished upper chest breath sounds on clinical examination. There was no evidence of focal neurologic signs or clinical evidence of NF-1 or Marfan syndrome on physical examination. The laboratory test results were within normal ranges.

Access this article online

www.ijss-sn.com

Month of Submission : 11-2022
Month of Peer Review : 12-2022
Month of Acceptance : 12-2022
Month of Publishing : 01-2023

Corresponding Author: Vignesh Gadupudi, A. C. S Medical College and Hospital, Chennai - 600 006, Tamil Nadu, India.

The patient underwent MR imaging. MR imaging confirmed the presence of thoracal vertebral segmentation anomalies and a large anterior meningocele herniating into the thoracic cavity through a bone defect at the level of T3-T4 vertebrae [Figure 1]. The cystic lesion was close to the trachea and the main vessels of the mediastinum, and it pushed the trachea to the left [Figure 2]. The patient had dorsal spine scoliosis and butterfly vertebrae at T1, T2, and T3 vertebral levels [Figure 3]. Sagittal T1 WI shows herniation of cord and CSF filled sac in anterior thoracic region through bony defect [Figure 4].

At the level of T3, the spinal cord was displaced anterior to the Meningocele's neck. There was no discernible change in parenchymal signal intensity or syrinx in the spinal cord. The meningocele was scheduled for surgical intervention with an anterior approach due to its size and location. However, the patient was lost to follow-up.

DISCUSSION

The herniation of meninges through bone defects or foramina to form a CSF-filled sac is referred to as a spinal meningocele. This is classified as either acquired or congenital, with acquired meningocele being a complication of laminectomy.[1] Neural tube defects (NTD) are common congenital disorders, with thoracic meningomyelocele accounting for 1–5% of all NTD.[2] Ectodermal cells proliferate near the midline of the embryo during normal embryogenesis in the 2nd week of pregnancy, forming the neural plate. On day 17, this plate invaginates, with adjacent lateral thickening forming the neural folds. The pathogenesis of spinal dysraphism is a failure of neural tube closure, which occurs in a bidirectional pattern beginning in the mid-cervical region and progressing cranially and caudally. According to this theory, the most common location of MMC is at the most cranial or most caudal ends; however, this theory fails to explain MMC

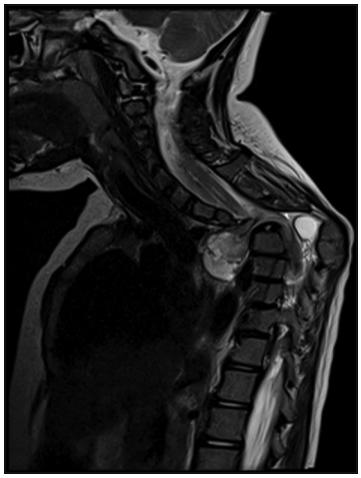

Figure 1: Sagittal T2 showing herniation of cord and CSF filled sac in anterior thoracic region through bony defect

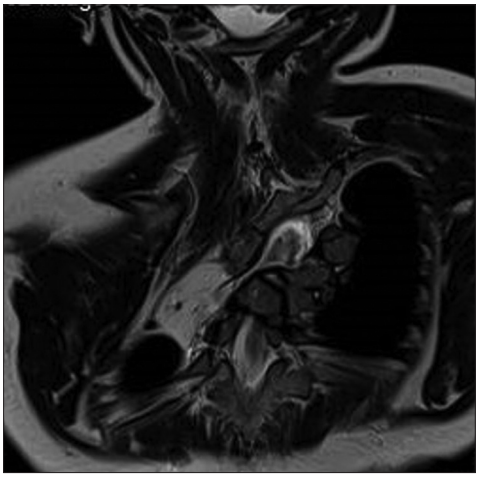

Figure 3: Coronal T2 WI showing scoliosis of dorsal spine and butterfly vertebrae of D1, D2, and D3

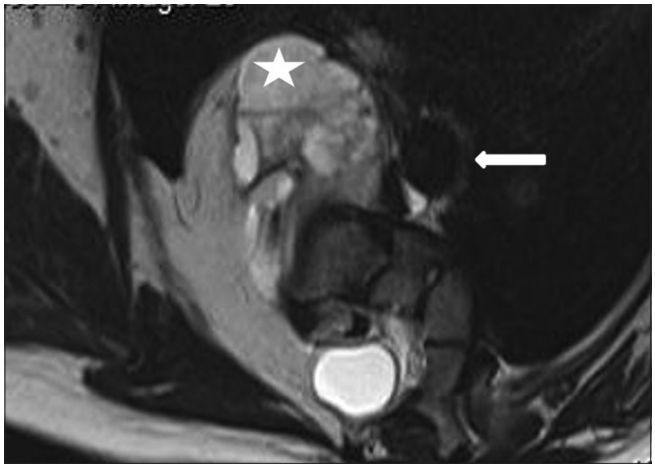

Figure 2: Axial T2 WI showing displacement of trachea (white arrow) to the left side by the meningomyelocele (asterix)

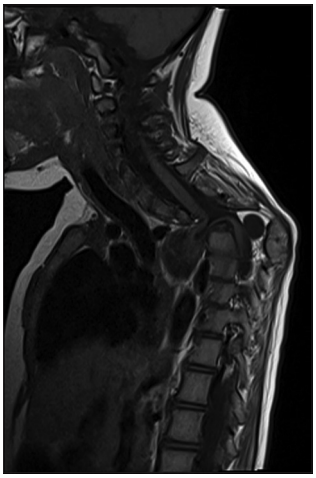

Figure 4: Sagittal T1 WI showing herniation of cord and CSF filled sac in anterior thoracic region through bony defect

in the thoracic region. Instead of forming a tube, the neural folds remain as a flat plate of tissue known as the neural placode. These defects can occur in any part of the vertebral column, with posterior defects being more common than anterior. Mesoderm development around the neural tube is incomplete in anterior meningocele.[1] A thoracic meningocele's clinical manifestations are closely related to its size and relationship to surrounding structures. Back pain, paraparesis from spinal cord injury, or shortness of breath, coughing, and palpitation caused by compression of the lung and mediastinal structures, as was the case for the patient presented here, are all possible symptoms. Even progressive hydrothorax caused by meningoceles rupture has been reported in the literature.[3] They are associated with less neurological deficits than their lumbosacral counterparts, but if left untreated due to tethering, they can progress to neurologic deficits.[2,4] In the presence of gross neurologic deficits in thoracic MMC, kyphosis occurs invariably due to unrestricted pull of the normally innervated proximal anterior abdominal and intercostal muscles, preventing tensionless closure of the defect and may necessitate kyphectomy, thereby improving lung endurance and functional capacity.

Intradural exploration with meticulous microsurgical release of the spinal cord through careful resection of all tethering bands has been suggested in those cases.[1] This case raises awareness of the thoracic MMC's distinctive presentation. A Cystoperitoneal shunt is the gold standard treatment for anterior dorsal meningocele; however, treatment options vary depending on the size of the lesion.[3,5] Laminectomy and duroplasty are preferred in smaller lesions of anterior dorsal meningocele, while thoracotomy can be planned in larger lesions.[2,4]

Cross-sectional imaging methods, such as computed tomography and magnetic resonance imaging (MRI), are crucial, regardless of the choice of treatment, not only for the diagnosis but also for the depiction of its relationships to surrounding structures and the exclusion of any additional potential accompanying lesions, such as a neuroma in the context of NF-1. The extension of the spinal canal and segmentation defects brought on by the thoracic meningocele in this case were detected by MR imaging, which also revealed the anatomic connections.

CONCLUSION

Spinal cord lesions can be diagnosed on MRI. MRI imaging with its excellent soft tissue contrast helps in accurately mapping the anatomy and enables surgical planning. The case report's goal is to raise awareness of the MMC's uncommon thoracic spine presentation and the possibility of scoliosis association with the lesion.

REFERENCES

1. Gutierrez FR, Woodard PK, Fleishman MJ. Normal anatomy and congenital anomalies of the spine and spinal cord. In: Osborn AG, Maack H, editors. Diagnostic Neuroradiology. 1st ed. St. Louis: Mosby; 1994. *p.* 785-819.
2. Von Koch CS, Glenn OA, Goldstein RB, Barkovich AJ. Fetal magnetic resonance imaging enhances detection of spinal cord anomalies in patients with sonographically detected bony anomalies of the spine. J Ultrasound Med 2005;24:781-9.
3. Mizuno J, Nakagawa H, Yamada T, Watabe T. Intrathoracic giant meningocele developing hydrothorax: **A** case report. J Spinal Disord Tech 2002;15:529-32.
4. Kumar J, Afsal M, Garg A. Imaging spectrum of spinal dysraphism on magnetic resonance: A pictorial review. World J Radiol 2017;9:178-90.
5. Tortori-Donati P, Rossi A, Cama A. Spinal dysraphism: A review of neuroradiological features with embryological correlations and proposal for a new classification. Neuroradiology 2000;42:471-91.

How to cite this article: Gadupudi V, Chaparala P, Vidavaluru AS. Isolated Anterior Thoracic Myelomeningocele: A Rare Case Report. Int J Sci Stud 2023;10(10):4-6.

Source of Support: Nil, **Conflicts of Interest:** None declared.