Am J Transl Res 2025;17(5):3546-3553
www.ajtr.org /ISSN:1943-8141/AJTR0161950

Case Report

Isolated true anterior thoracic meningocele associated with severe kyphoscoliosis: a case report

He-Cheng Zhang[1*], Quan-Shi[2*], Xin Zhang[3], Wei-Xin Li[4]

[1]Department of Orthopedics, Fengxian County People's Hospital, Fengxian, Jiangsu, China; [2]Department of Nosocomial Infection, Fengxian County People's Hospital, Fengxian, Jiangsu, China; [3]Department of Orthopedics, Qinghai Provincial People's Hospital, Xining, Qinghai, China; [4]Department of Neurosurgery, Tangdu Hospital, Xi'an, Shanxi, China. [*]Equal contributors.

Received November 12, 2024; Accepted April 23, 2025; Epub May 15, 2025; Published May 30, 2025

Abstract: Introduction: Spinal meningoceles are congenital anomalies characterized by meningeal herniation through vertebral defects, most commonly occurring in the lumbosacral region. Intrathoracic meningoceles are rare and are typically associated with neurofibromatosis type 1 (NF-1). True anterior thoracic meningoceles unassociated with NF-1 represent exceptionally rare clinical entities. Case summary: This is a retrospective case report. We report a 15-year-old female with severe kyphoscoliosis and a non-NF-1-associated anterior thoracic meningocele who presented with progressive spastic paraparesis. Imaging examinations revealed thoracolumbar scoliosis with 100-degree kyphotic angulation, midline anterior spina bifida at T7, and a well-defined cystic structure exhibiting hypointense T1 and hyperintense T2 signals (measuring 5.7×4.5 cm) anterior to the T7-T8 vertebral bodies. Surgical intervention included posterior spinal laminar decompression, T7 vertebral osteotomy, microscopic dural sac reduction, and stabilization using a pedicle screw system with autologous bone grafting. The patient was discharged without any complications 12 days after surgery, and at the 12-month follow-up, the patient achieved ambulation with a walker and leg brace. Conclusion: This case underscores the necessity of tailored surgical strategies for anterior thoracic meningoceles complicated by severe spinal deformities. The integration of microsurgical techniques and 3D-printed anatomical modeling may optimize procedural safety and functional outcomes.

Keywords: Anterior thoracic meningocele, kyphoscoliosis, paraparesis, surgery, case report

Introduction

Spinal meningocele is a congenital deformation affecting the central nervous system, and deficiencies in folic acid and zinc during fetal development have been shown to be the cause of this disease [1]. More than 80% of spinal meningoceles are located posteriorly in the lumbosacral area. Anterior meningoceles are rarer and are generally described in the thoracic and sacral regions [2]. Spinal meningoceles clinically manifest as dorsolumbar masses accompanied by neurological deficits (e.g., lower extremity paresis, dermatomal sensory impairment) and neurogenic bladder/rectal dysfunction. The association between meningoceles and kyphosis stems from congenital vertebral defects disrupting spinal alignment. Mechanical stress at the defect site exacerbates anterior spinal column collapse, creating a vicious cycle of deformity progression.

An intrathoracic meningocele, first documented by Pohl [3] in 1933, represents a very rare variant. Most reported thoracic meningoceles originate laterally through expanded intervertebral foramina and are not strictly anterior [4]. In contrast, the thoracic meningocele described in our case arose from a broad midline bony defect of the vertebral bodies, not through an enlarged foramen. This unique anatomical feature distinguishes it as a true anterior thoracic meningocele. Andrade et al. [5] identified 134 intrathoracic meningocele cases reported before 1992, with 69% associated with neurofibromatosis type 1 (NF-1) and only 22% classified as isolated. Current treatments range from conservative monitoring for asymptomatic patients to surgical repair via posterior laminectomy or combined anterior-posterior approaches, depending on the lesion size and spinal stability [6]. Clinical outcomes demonstrate marked

https://doi.org/10.62347/SMEE7102

neurological improvement in 71.4% of patients following surgical repair, whereas untreated progressive lesions may lead to irreversible paralysis secondary to spinal cord compression and ischemic injury [7].

Only two cases of isolated true anterior thoracic meningocele combined with kyphoscoliosis have been reported worldwide according to a PubMed search, and no cases have been reported in China [8, 11]. Here, we report a third case. Compared with the previously reported cases, we also used more cutting-edge treatment options, such as microscopy and 3D printing, which greatly reduced the risk of the corrective surgery. We provide this case as a reference for clinical treatment.

Case presentation

Chief complaints

A 15-year-old girl presented to the Emergency Department of our hospital complaining of worsening lower limb weakness.

History of present illness

The patient's symptoms started four years prior, with lower limb weakness, which had worsened in the last 24 h.

History of past illness

Four years ago, the patient gradually developed lower limb weakness and was diagnosed with scoliosis after visiting a local hospital. She was not receiving any treatment for economic reasons.

Personal and family history

There was no family history of spinal deformity, and she had no stigmata of neurofibromatosis or Marfan syndrome.

Physical examination

No "café au lait" spots, abnormal hair, or lumps were found after examination of her back. The thoracic spine was deformed to the left, the right shoulder was slightly higher, the left waist skin was wrinkled, and the left iliac crest was slightly higher. A razorback deformity was visible when bending over. The spinous processes of each vertebral body had no tenderness, and the thoracolumbar spine was mobile.

Both lower limbs appeared abnormal. The iliopsoas muscle strength of both lower limbs was Grade II, the rectus femoris and tibialis anterior muscles were Grade I, and the dorsi extensor and plantar flexor muscle strengths were Grade 0. Muscular tension in both lower limbs was increased, with a negative abdominal wall reflex, negative bilateral patellar clonus and ankle clonus, and a weakened bilateral knee reflex and Achilles tendon reflex. Bilateral Hoffman signs were negative, and the remaining bilateral pathological signs were positive. The positive bilateral pathological signs indicated upper motor neuron injury, likely caused by chronic spinal cord compression from both the anterior meningocele and severe kyphoscoliosis.

Imaging examinations

Radiographs revealed kyphoscoliosis with a kyphotic curvature of more than 100 degrees around the cervicothoracic junction from T1 to T12, while the scoliosis had a Cobb angle of 36 degrees from T1 to T12 (**Figure 1**). Three-dimensional CT revealed T6-T8 vertebral and lamina fusion and vertebral spina bifida in front of T7 (**Figure 2**). Magnetic resonance imaging (MRI) revealed the presence of multiple thoracic vertebral anomalies, and the anterior meningocele had herniated into the thoracic cavity through a bone defect at the level of the T7 vertebra and was approximately 5.7×4.5 cm in size. The spinal cord was displaced anteriorly at the T7 level and draped over the superior aspect of the meningocele sac (**Figure 3**).

Final diagnosis

The patient was diagnosed with anterior thoracic meningocele, kyphoscoliosis, T7 vertebral spina bifida, and paraparesis.

Treatment

General anesthesia was applied, and the patient was placed in the prone position. A T2 to T11 segment incision was made in the posterior midline of the spine, exposing the transverse process of the affected side and showing the left side curvature and kyphosis deformity of the thoracic spine, T5 to T8 spinous processes, and lamina fusion. The pedicle screws were first sequentially implanted in the bilateral T3, T4, T5, T9, T10, and T11 pedicles from the per-

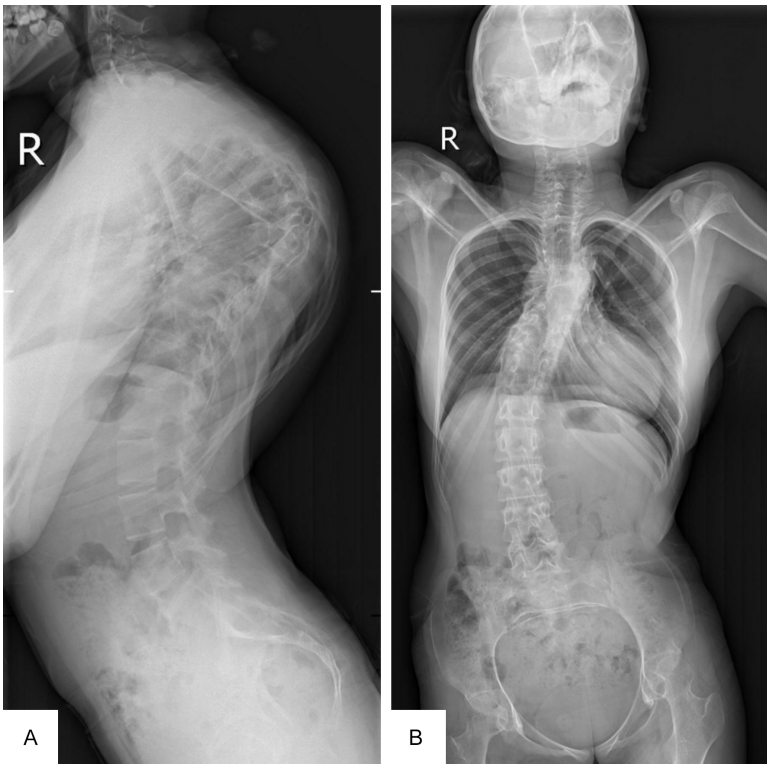

Figure 1. A: Lateral position X-ray image showing kyphosis curvature greater than 100 degrees from T1 to T12. B: Anteroposterior position X-ray showing that the scoliosis had a Cobb angle of 36 degrees from T1 to T12.

spective of the C-arm machine. The bilateral fixing rod was fixed temporarily to avoid damage to the spinal cord and the nerve root. Decompressive laminectomy was performed intraoperatively, with the right rib head and transverse process at T6, T7, and T8, and the whole lamina was excised. The dura mater had no tension, and epidural fat was present. Entering along the T6 and T7 pedicles, part of the vertebral body was excised to reveal the meningocele.

A microscope was mounted and the dura was microscopically cut by a neurosurgeon, and the spinal cord, hernia sac, and exit were explored with a nerve root dissector. Part of the hernia sac was gray, and dark green fluid could be drawn from the sac. The lateral sac fluid was clear cerebrospinal fluid. An intestinal cyst was suspected. Continuing to peel forward along the wall of the hernia sac, it was found that the anterior wall of the spinal cord had been pushed into the thoracic cavity by a cyst. The wall of the sac was thick and strongly adhered to the spinal cord. Owing to concern that the internal volume of the spinal canal would be insufficient for complete reduction, only partial reduction was performed, and the remaining part was suspended with a Mousse thread to the rear.

Further treatment of the T7 vertebral spina bifida and kyphosis was performed as follows. A wedge-shaped osteotomy (SPO) was performed between the left pedicles of T6 and T7 to cut the vertebral body forward, limb elevation and cantilever reduction were used, and the lever-assisted reset technique was used to correct the kyphotic deformity. The osteotomy surface was closed, autogenous bone was implanted, bilateral rods were fixed, and horizontal connecting rods were placed for connection. The T7 vertebral spina bifida was managed by SPO between the T6 and T7 pedicles to reconstruct the anterior spinal column, which was combined with autologous bone grafting and pedicle screw fixation to restore structural integrity. Kyphosis correction was achieved through T6-T7 SPO and the lever-assisted reset technique, which reduced the Cobb angle from >100° to 45°. The decision against aggressive orthopedic surgery was based on the high risk of spinal cord ischemia during acute angular correction [7]. The surgical field was carefully irrigated, the bilateral lamina cortex was removed, and autologous bone particles were implanted for vertebral fusion. Then, an indwelling drainage tube was placed, and the incision was closed (**Figure 4**).

The total operating time was 360 min, and the volume of blood lost was 1,400 mL. During surgery, 900 mL of red blood cells and 600 mL of plasma were infused.

Outcome and follow-up

Postoperatively, her paralysis remained complete. Both lower limbs remained abnormal on physical examination, her muscle strength did not improve compared with that preoperatively, and the bilateral pathological signs were posi-

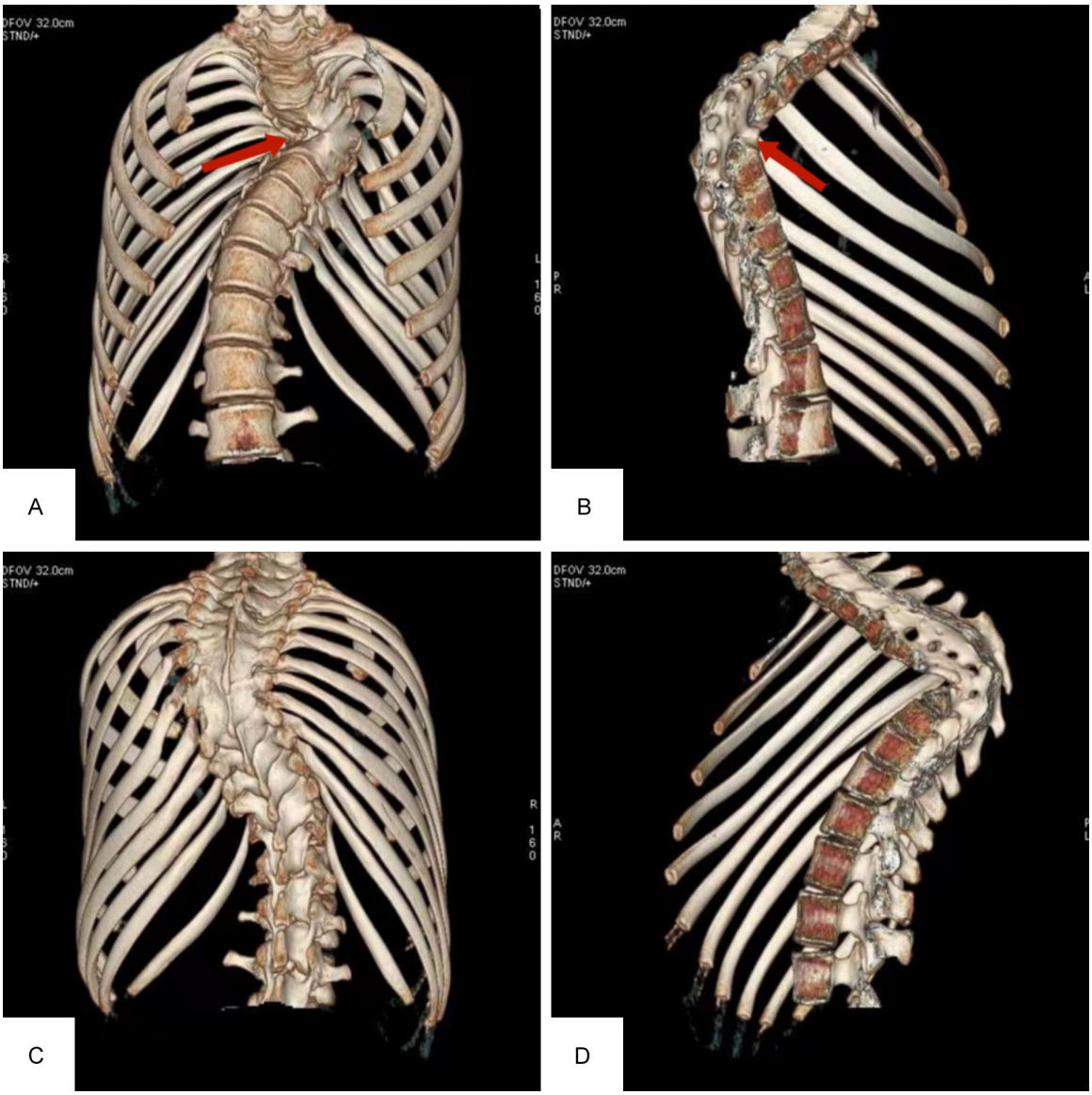

Figure 2. Three-dimensional CT revealed that the thoracic scoliosis was deformed with angled kyphosis; the T7 anterior vertebral body spina bifida (red arrow. A/B); the T4-T8 vertebral body morphology was abnormal, the intervertebral space fusion disappeared (C), and the spinal canal was unevenly expanded (D).

tive. Postoperatively, both lower limbs remained abnormal due to chronic spinal cord injury from long-term compression, despite decompression and deformity correction. After the patient's condition stabilized, she was discharged from the hospital and referred to the rehabilitation hospital for neurological rehabilitation treatment. Recently, a follow-up was conducted one year after the operation, and the patient was in good condition and could walk with the help of a walker and leg brace. One year after the operation, plain X-ray imaging revealed that the internal fixation device was well positioned without breakage (**Figure 5**). MRI revealed no neural tissue in the thoracic hernia sac contents (**Figure 6**).

Results

Anterior thoracic meningocele is a rare disease. For patients with meningoceles combined with kyphoscoliosis, severe progressive kyphosis causes instability around the apex of the kyphoscoliosis and spinal cord compression. The formulation of the surgical protocol should be individualized according to the location, size, and presence of compression symptoms. Intraoperative microscopic separation of severely

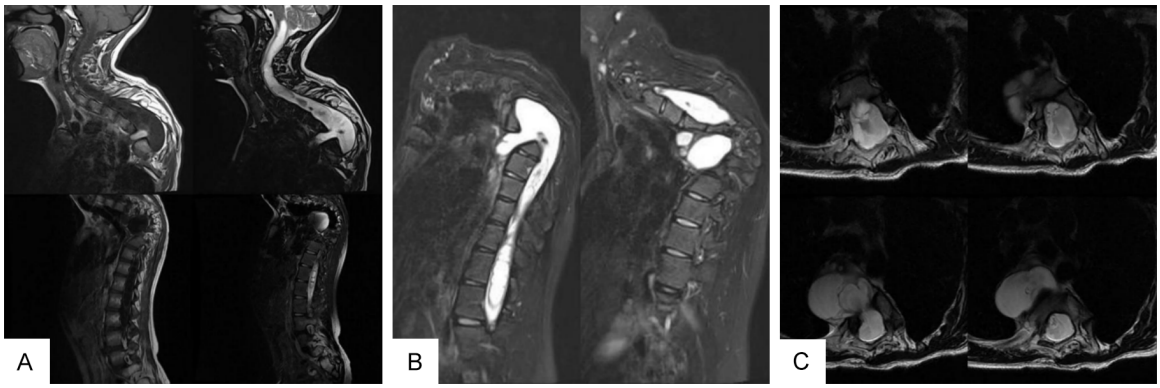

Figure 3. MRI shows cystic expansion of the T4-T8 vertebral lumen and a bag-like long T1 and long T2 signal shadow in front of the T7-T8 vertebral level, corresponding to a change in the vertebral spina bifida (A/B). A meningocele was considered above. The internal signal was not uniform, and a few flocculent spinal cord shadows were observed (C).

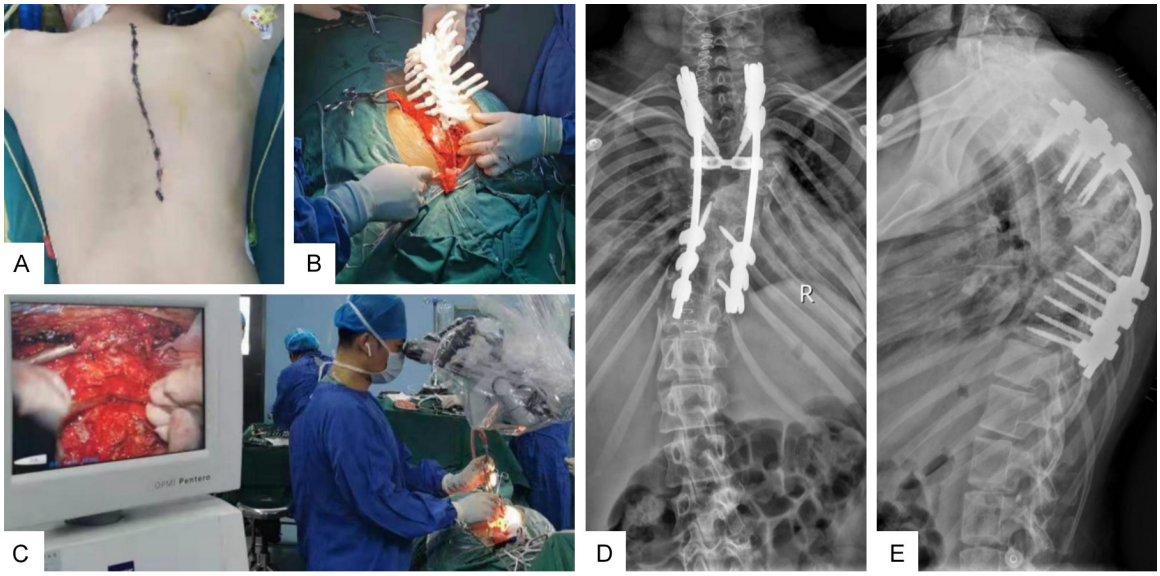

Figure 4. A: The patient was in a prone position and was marked with a surgical incision from T2 to T11 at the midline of the posterior spine. B: Preoperative 3D printing based on the patient's MRI and CT data to guide pedicle screw placement and wedge osteotomy. C: The dura was cut microscopically by a neurosurgeon, and the spinal cord, hernia sac, and exit were explored with a nerve root dissector. D/E: X-ray taken immediately after the operation showing that T3, T4, T5, T9, T10, and T11 bilateral pedicle screws were implanted, fixed with bilateral rods and connected by horizontal connecting rods.

adherent hernial sac contents can significantly reduce the risk of spinal cord injury. For severe spinal deformities, intraoperative pedicle screw placement and wedge osteotomy can be guided by a 3D spine model. Postoperative rehabilitation and brace therapy can also provide the best assistance for neurological recovery in such patients.

Discussion

Anterior meningoceles are a herniation of the meninges through the foramina or a defect in the vertebral column and are commonly accompanied by NF-1. The anterior thoracic meningocele described in this case was associated with congenital spinal anomalies, with the vertebral spina bifida visible in front of T7, through which the meningocele protruded. Tsou et al. [9] proposed that midline sagittal defects in the vertebral bodies develop due to midline adhesions between the endoderm and ectoderm layers of the embryonic disc. As the notochord migrates cephalad between these layers, it must fork at the point of endoderm-ectoderm adhesion,

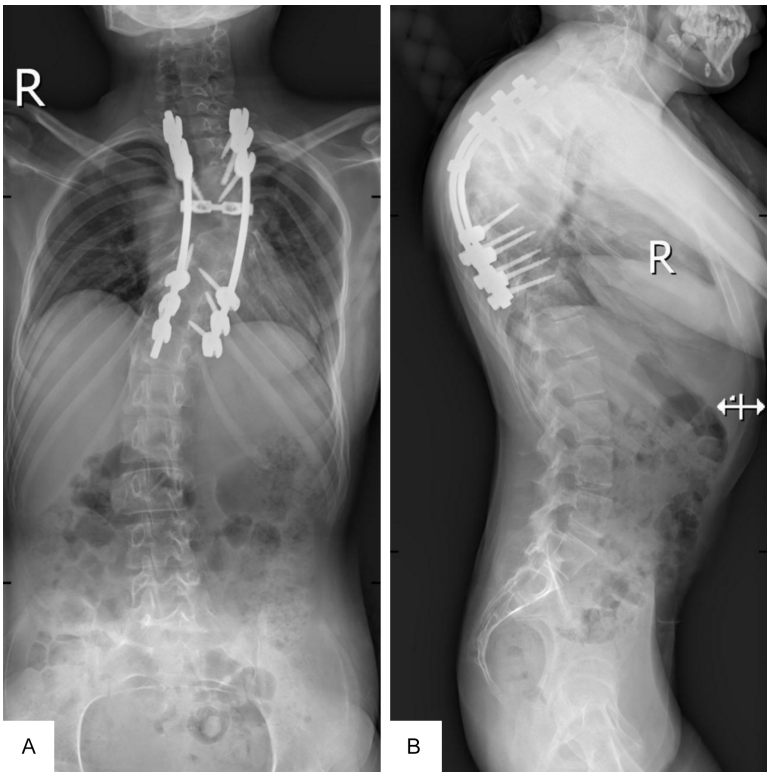

Figure 5. A/B: Anteroposterior and lateral position X-rays showing that the spinal internal fixation device was well positioned without breakage.

resulting in a midline defect in the vertebra. Myelomeningocele elements may herniate anteriorly to form anterior myelomeningoceles in the thoracic spine. Differential diagnoses include posterior mediastinal masses such as neurofibromas, neuroblastomas, and cystic hygromas, which require contrast-enhanced MRI and histopathological confirmation for definitive diagnosis [10].

In our case, the T7 anterior vertebral defect and severe kyphosis were probably responsible for the formation of an anterior meningocele. In contrast, her paraparesis was the result of a severe, progressive spinal deformity. There is a consensus that intrathoracic meningoceles rarely require resection, and an operation to treat an intrathoracic meningocele should be considered only when there are complaints of pain, neurological or pulmonary signs, or compression of the trachea or esophagus [11]. In our case, the patient had already developed paraplegia.

The surgical plan is closely related to the lesion size. For small or medium-sized meningoceles, laminectomy and intradural repair of the cyst are recommended. The advantage is that thoracotomy is avoided, and spine stability is more conservative because the posterior ligament remains intact. The posterior approach is generally assumed to be inadequate for larger lesions, and transthoracic access is indicated. It offers a more extensive surgical field with only a slight risk of damaging the spinal cord [2, 5].

In recent years, with the rise of microscope technology, the adhered dura and spinal cord can be clearly separated under a microscope, significantly reducing the risk of spinal cord injury [12]. Therefore, we recommend that for small or medium-sized meningoceles with acceptable surgical risk, adequate definitive treatment should primarily be performed through a posterior approach under the microscope. Considering that changes in cerebrospinal fluid dynamics may lead to serious neurological complications after surgery and that our patient did not have pulmonary signs or tracheal or esophageal compression symptoms, we did not perform hernia sac resection and instead returned the dura mater adhering to the spinal cord to the spinal canal, and the remaining part was fixed to the rear.

Severe progressive kyphosis causes instability around the apex of the kyphoscoliosis and spinal cord compression, and laminectomy further increases the risk of spinal instability [13]. Therefore, our patient underwent wedge osteotomy during surgery for the meningoceles, and multiple reduction techniques, along with spinal bone graft fusion and the placement of a pedicle screw system, corrected the kyphotic deformities.

Notably, a 3D spine model produced from the patient's preoperative CT data can be used to formulate the surgical plan before the operation and can be repeatedly viewed on the operating table after low-temperature disinfection

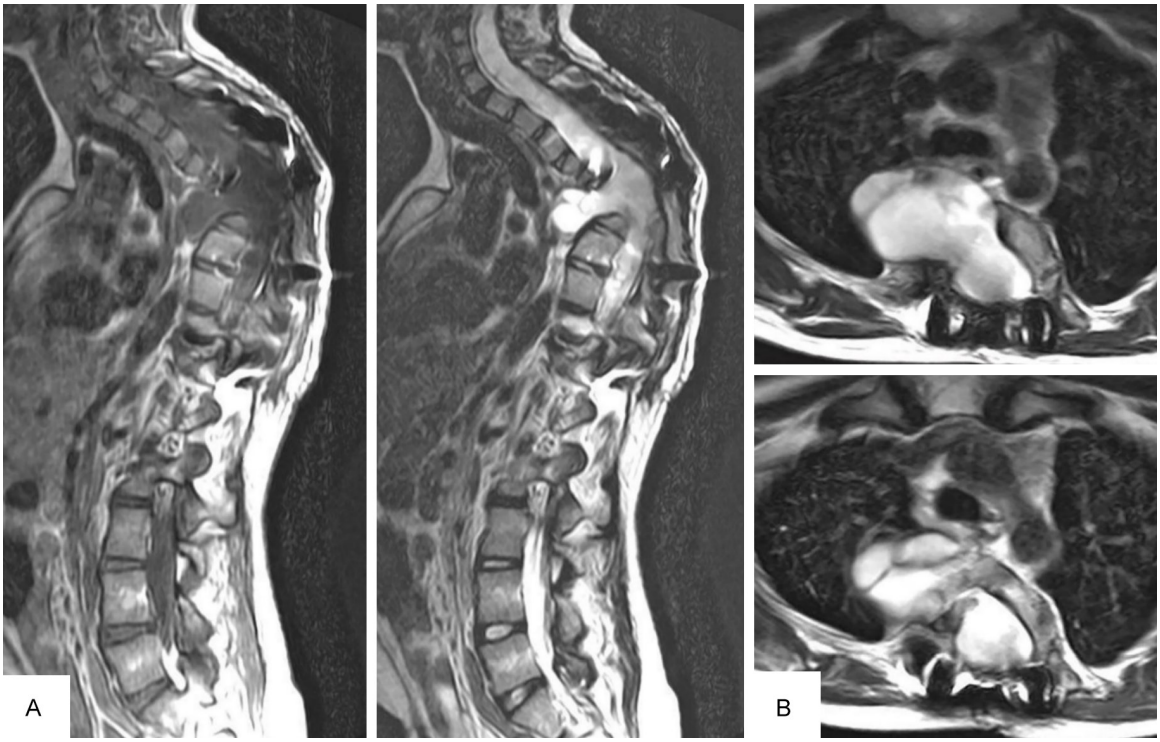

Figure 6. MRI revealed a bag-like long T1 and long T2 signal shadow in front of the T7-T8 vertebral body, with a range of approximately 5.76×4.05 cm (A). Compared with the preoperative signal, the signal inside the bulging meninges was uniform, and no clear spinal cord signal shadow was found (A/B). The lesion locally communicated with the adjacent spinal canal, and the spinal canal was dilated at the level of the T4-T8 vertebral body (A).

to guide pedicle screw placement and wedge osteotomy.

We suggest that for patients with paraplegic symptoms, we can perform preoperative high-dose hormone shock therapy and request absolute bed rest to reduce the risk of further spinal cord injury. Postoperatively, patients should wear chest and leg braces and perform rehabilitation exercises to promote neurological recovery.

Acknowledgements

We would like to thank the patient and her family for their participation, along with Wei-Xin Li, the neurosurgeon at Xi'an Tangdu Hospital, for providing free medical and technical support to the patient.

Disclosure of conflict of interest

None.

Address correspondence to: Xin Zhang, Department of Orthopedics, Qinghai Provincial People's Hospital, Xining 810007, Qinghai, China. E-mail: 13897479287@163.com

References

[1] Copp AJ, Adzick NS, Chitty LS, Fletcher JM, Holmbeck GN and Shaw GM. Spina bifida. Nat Rev Dis Primers 2015; 1: 15007.

[2] Chen HC, Chang PH, Jhang SW and Wang BY. Thoracoscopic plication for a huge thoracic meningocele in a patient with neurofibromatosis. J Cardiothorac Surg 2014; 9: 85.

[3] Phol R. Meningocele in brustraum unter dem bilde eines intrathoraken brandschatten (in German). Rontgenpraxis 1933; 5: 747-9.

[4] Maldonado RG, Extremera BG, Alegre V and Vara Thorbeck R. Intrathoracic meningocele presenting as a mediastinal mass lesion. Case report. Zentralbl Neurochir 1992; 53: 11-14.

[5] de Andrade GC, Braga OP, Hisatugo MK, de Paiva Neto MA, Succi E and Braga FM. Giant intrathoracic meningoceles associated with cutaneous neurofibromatosis type I: case report. Arq Neuropsiquiatr 2003; 61: 677-681.

[6] North RB, Kidd DH and Wang H. Occult, bilateral anterior sacral and intrasacral meningeal and perineurial cysts: case report and review of the literature. Neurosurgery 1990; 27: 981-986.

[7] Venanzi MS, Martini G, Rossi A, Piatelli G and Pavanello M. Intrasacral meningoceles: clini-

cal presentation, surgical management, and postoperative outcome: the Giannina Gaslini Hospital's experience. Neurochirurgie 2023; 69: 101466.
[8] Jaffray D and O'Brien JP. A true anterior thoracic meningocele associated with a congenital kyphoscoliosis. J Pediatr Orthop 1985; 5: 717-719.
[9] Tsou PM, Yau A and Hodgson AR. Embryogenesis and prenatal development of congenital vertebral anomalies and their classification. Clin Orthop Relat Res 1980; 211-231.
[10] Zuo T, Ni Z, Zhou M, Huang C, Tang Z and Yang Z. Large intrathoracic meningocele in a patient with neurofibromatosis type 1. Asian Cardiovasc Thorac Ann 2014; 22: 219-221.
[11] Oner AY, Uzun M, Tokgöz N and Tali ET. Isolated true anterior thoracic meningocele. AJNR Am J Neuroradiol 2004; 25: 1828-1830.
[12] Ma L and Fei B. Comprehensive review of surgical microscopes: technology development and medical applications. J Biomed Opt 2021; 26: 010901.
[13] Zhou Y, Liu CZ, Zhang SY, Wang HY, Nath Varma S, Cao LQ, Hou TT, Li X and Yao BJ. Giant schwannoma of thoracic vertebra: a case report. World J Clin Cases 2021; 9: 11448-11456.