Intrathoracic meningocele associated with neurofibromatosis Type 1 and a novel technique for surgical repair: case report

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Neurofibromatosis Type 1 (NF1) is an autosomal dominant neurocutaneous disorder that can have associated spinal abnormalities related to both bone and dural dysplasia. Thoracic meningoceles are one spine anomaly associated with NF1, although they are a fairly uncommon pathology. Surgical techniques to treat these meningoceles, usually undertaken only when the patient is symptomatic, are targeted at decreasing the size of the protrusion and improving lung capacity. Surgical interventions discussed in the literature include shunting the pseudomeningocele, primary repair with laminectomy, thoracoscopic plication, and reinforcement of the closure with cement, muscle, or fascia. Authors here report the case of a 43-year-old woman with NF1 with worsening pulmonary function tests and in whom shunting of the pseudomeningocele failed. Subsequently, a posterolateral thoracotomy was performed. The dura mater was reconstructed and primarily closed. On this closure a Gore-Tex soft-tissue patch was placed along with polypropylene mesh and Evicel fibrin sealant, followed by titanium mesh. At the end of the procedure, a chest tube was left in place and therapeutic pneumoperitoneum was performed to decrease the dead space as the lung did not fully expand with positive-pressure ventilation. The patient’s pulmonary function tests improved after the procedure.

Thoracic meningoceles are uncommon and difficult pathologies to treat surgically. Although shunting is arguably the least invasive surgical option, it can fail in some patients. When it does fail, there are other options that require a multidisciplinary approach and careful attention to the dural closure and reinforcing layers.

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NEUROFIBROMATOSIS Type 1 (NF1) is an autosomal dominant neurocutaneous disorder with complete penetrance, affecting 1 in 3000 individuals among all ethnic groups. Spinal abnormalities related to both bone and dural dysplasia are seen specifically in NF1, and not in NF2. The incidence of spinal deformities in NF1 ranges from 10% to 60%, with scoliosis as the most common spinal manifestation. The remainder involve both soft-tissue and osseous complications and include pedicle erosion and enlargement of the intervertebral foramina. Importantly, the dura mater itself is also abnormal.

Dural ectasia refers to widening or outpouching of the dural sac, which can manifest as meningoceles herniating through enlarged intervertebral foramina, scalloping of the vertebral bodies, or erosion of other osseous spinal elements such as the pedicles. The majority of dural ectasias are seen in NF1, but they are also strongly associated with Marfan syndrome. Other associated conditions include ankylosing spondylitis and achondroplasia, although these may have different mechanisms. The true pathogenesis behind the dural and bony abnormalities in NF is not known; however, it is thought that underlying mesodermal dysplasia affects both the osseous structures and the dura. It is also possible that the dural expansion puts stress on surrounding bone, contributing to the scalloping and thinning of the pedicles.

Thoracic meningoceles associated with NF1 were first described in 1933 by Phol. Overall, it is an uncommon pathology, but 69% of thoracic meningoceles are associated with NF1. In the thorax, lateral meningoceles are more common because the paravertebral muscles are relatively weak and the pressure gradient between the cerebrospinal fluid and the thoracic cavity is lower.

ABBREVIATIONS CSF = cerebrospinal fluid; NF1 = neurofibromatosis Type 1; PEEP = positive end-expiratory pressure.


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fluid (CSF) space and the pleural space is higher.⁴ Often there is an associated kyphoscoliotic deformity of the thoracic spine.⁵,⁶ Pseudomeningoceles have been reported in the cervical and lumbar spine as well; however, they are fairly uncommon.⁹,¹²

Meningoceles can be asymptomatic or present with neurological or pulmonary findings. They can cause paraparesis, pain from compression of the spinal cord, or low-pressure headaches. There may also be compression of the lung cavity or mediastinal structures, which can cause cough, dyspnea, and palpitations.¹⁷ Surgical techniques to treat these meningoceles, usually undertaken only when the patient is symptomatic, are targeted at decreasing the size of the protrusion and improving lung capacity.

Case Report
History and Examination

A 43-year-old woman was initially diagnosed with NF1 at the age of 9 after she had presented with scoliosis. She underwent spinal fusion with Harrington rod placement at the age of 12 and was lost to follow-up. She presented to us with back pain and dyspnea that had been progressive for the past 2 years. On examination, she was neurologically intact with scoliosis. Significant restrictive lung disease was present, with pulmonary function tests at 38% of age-adjusted normal. Magnetic resonance imaging of the spine and CT scanning of the chest showed a large right thoracic meningocele, along with dystrophic changes and scalloping of the midthoracic spine from T-8 to T-11 (Fig. 1).

First Treatment

As initial treatment, we placed a cystoperitoneal shunt with a programmable Strata valve (Medtronic) adjusted to a pressure of 0.5. Because of low-pressure headaches, the valve was eventually adjusted to 2.0. The patient also seemed to have neck pain as a symptom of overdrainage that resolved with increasing the shunt valve setting. The thoracic meningocele did not change in size, pulmonary function tests did not improve, and the patient did not report relief of her dyspnea. Two large-volume taps of the shunt at 1 L and 1.2 L on 2 separate occasions, as well as changing the pressure setting of the valve down to 1.0, were attempted without improvement in the patient’s symptoms. Immediately after these taps, the volume of the pseudomeningocele would be smaller on imaging but the fluid would reaccumulate (Fig. 2).

Second Treatment

Since the patient did not improve, a right posterolateral thoracotomy with resection of the meningocele and reconstruction of the dural sac was performed with the assistance of a thoracic surgeon. At the seventh intercostal space, the large dural sac was drained and found to contain nearly 2 L of CSF. The spinal cord was identified prior to resection of the sac (Fig. 3). The dura was closed primarily using silk sutures, and a polypropylene mesh was used to reinforce the repair. Evicel (Ethicon Inc.), a fibrin-based dural sealant, was also applied. Valsalva maneuvers were used to confirm the absence of a CSF leak, and closure was performed with the assistance of the thoracic surgeons and a chest tube was left in place. Lumbar drainage was used postoperatively for 7 days, and the patient was kept on bed rest.

Third Treatment

Because of persistent filling of the pseudomeningocele, the patient was taken back to the operating room for repeat repair of the meningocele. Endotracheal intubation was
achieved with a double lumen tube to allow for single lung ventilation when repairing the pseudomeningocele. The thoracotomy was reopened, and the dura again closed primarily. A Gore-Tex soft-tissue patch (Gore Medical) was placed over the repair and Evicel was applied. Over this, polypropylene mesh was placed and Evicel was reapplied. Over this, a piece of titanium mesh was secured with a cranial plating system (DePuy Synthes CMF system) and Evicel was reapplied. Initially to promote expansion of the lung, a positive end-expiratory pressure (PEEP) of 10 cm H2O was used with sustained high peak pressures. Although this resulted in some reexpansion, the dead space was not resolved and therefore therapeutic pneumoperitoneum was performed. One of the problems in a hydropneumothorax is that there is a larger pleural space because there is a reduction in lung volume in relation to chest cavity volume. The introduction of air into the peritoneal cavity displaces the diaphragm cephalad and decreases the volume of the chest cavity relative to lung volume.19

To perform the pneumoperitoneum, a purse-string suture was placed in the diaphragm. The diaphragm was then opened, and a 14-Fr red rubber catheter was placed into the peritoneum. The pneumoperitoneum was created by insufflating the abdomen with 2.5 L of air and monitoring airway pressures, blood pressure, and urine output. The defect in the diaphragm was sutured shut. Two chest tubes and a lumbar drain were placed.

Postoperative Course

The patient remained intubated on a PEEP of 10 cm H2O for 24 hours to promote lung reexpansion. Her chest tubes were weaned over 10 days, and the lumbar drain was removed on the 11th day after surgery. She recovered well, and postoperative pulmonary function tests showed an improvement to 50% of age-adjusted normal. Imaging 1 year after her procedure showed a decrease in the size of the pseudomeningocele (Fig. 4).

Discussion

Surgical intervention is indicated when a meningocele has a rapid rate of growth or when a patient is symptomatic due to compression of surrounding structures.21 The earliest surgical treatments involved posterior laminectomy with direct repair of the meningocele. However, laminectomy is contraindicated for severe kyphoscoliosis as it can accelerate progression of spinal deformity.19 Some suggest that if scoliosis is present, the patient may need extensive posterior fusion and anterior stabilization.

There have also been reports on the placement of cystoperitoneal and lumboperitoneal shunts using fixed pressure24 and programmable valves.20 However, this is not always a definitive treatment, and open surgery should be considered if there is no clinical improvement, as demonstrated in our patient. It is also possible that a patient presenting with low-pressure headaches will not have improvement in the headaches with a shunt.

When there is a large meningocele, thoracotomy is preferred. Watertight closure of the dura in these cases is difficult and postoperative fistulas can occur.16 There have been reports of closures that have been reinforced with cyanoacrylate cement, muscle, or fascia.20 As demonstrated in our patient, Gore-Tex or mesh may be another option for reinforcing the closure. It is unlikely that primary repair alone would be successful in these patients because of the inherent abnormalities in the dura; however, thoracoscopic plication has been successful.1

A posterolateral extradural approach has some advantages over an open thoracotomy. It does not require chest tube drainage, which can be advantageous as the dural repair may not be completely watertight. This technique may not be suitable for very large intrathoracic meningoceles as the resected dural sac may not be able to be removed without the spread or resection of the ribs, particu-
larly in a case in which the meningocele involves several vertebral levels.

Conclusions
Thoracic meningoceles are uncommon and a difficult pathology to treat surgically. Although shunting is arguably the least invasive surgical option, it can fail in some patients. When it does fail, there are more invasive options that require a multidisciplinary approach and careful attention to the dural closure and reinforcing layers. Here, we have presented a successful result in a patient in whom initial shunt placement had failed.

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Conception and design: Das. Analysis and interpretation of data: Goyal. Drafting the article: all authors. Critically revising the article: Das, Hunt. Reviewed submitted version of manuscript: Das. Approved the final version of the manuscript on behalf of all authors: Das. Study supervision: Hunt.

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