



Thoracic Meningoceles in NF1: Case Report and Considerations for Surgical Management

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Abstract

Thoracic meningoceles are rare entities seen in association with Neurofibromatosis type 1 (NF1). Although they uncommonly require resection, surgical techniques and postoperative management for these patients are highly varied and complex. Here, we report our experience with the first successful thoracotomy performed from a posterior-only approach for resection of a large thoracic meningocele. Additionally, we describe surgical and medical considerations for intra operative and postoperative management as well as potential complications.

Introduction

Neurofibromatosis Type 1 (NF1) is a genetic disorder of the neurofibromin gene that results in dysplasias of the skin, skeletal, and nervous systems [1-3]. Skeletal dysplasias in NF1 involve spinal deformities, including scoliosis, vertebral body scalloping, vertebral body wedging, vertebral rotation, and enlarged intervertebral foramina. Other dystrophic features include rib penciling, transverse process spindling, dural ectasia, and paraspinal tumors [4-7].

Defects of the bony spine can lead to meningoceles, which are often associated with disorders such as NF1, Marfan's syndrome, ankylosing spondylitis, and achondroplasia [8,2]. In the absence of an underlying connective tissue disorder, meningoceles can occur secondary to trauma or laminectomy surgeries [5,7]. When present, spinal meningoceles are usually lumbosacral 12 and rarely in the thoracic spine. However, 60% to 85% of the thoracic meningoceles in the literature have occurred in NF1 patients [2,9,10]. The pathophysiology of a meningocele is hypothesized to involve a focal herniation of the thecal sac through the enlarged intervertebral foramina. There is significant pressure difference between the Cerebrospinal Fluid (CSF) and pleural space in the thoracic spine, which is exaggerated by negative intrapleural pressure during inspiration [9,5-7,11]. Meningoceles can subsequently erode adjacent bone and create a larger defect [6].

Patients with meningoceles are typically asymptomatic but can present with dyspnea, coughing, or back pain [5,7]. When symptomatic, surgical interventions primarily aim to reduce mass effect and improve lung capacity. Case reports of surgical options include CSF diversion with cystoperitoneal shunting or direct drainage and excision of the meningocele [12,2,10]. Approaches to exposure depend on the size of the meningocele and degree of spinal deformity.

Here, we report the first posterior-only approach for resection of a thoracic meningocele and the associated medical and surgical considerations.

Case Presentation

History and examination

The patient is a 50-year-old male with NF1 with characteristic skin lesions and kyphoscoliosis, corrected with Harrington rod fusion. He was referred to neurosurgery when workup for back pain and shortness of breath revealed a left chest lesion associated with spinal deformities. He otherwise had an intact exam. CT Chest and MRI Thoracic spine (Figure 1) revealed a large cystic lesion, contiguous with the thecal sac that projected from multiple thoracic foramina.

Operation

The patient was taken to the operating room where a Lumbar Drain (LD) was placed. Via a vertical midline incision, he then underwent posterior left T6-11 hemilaminectomies,

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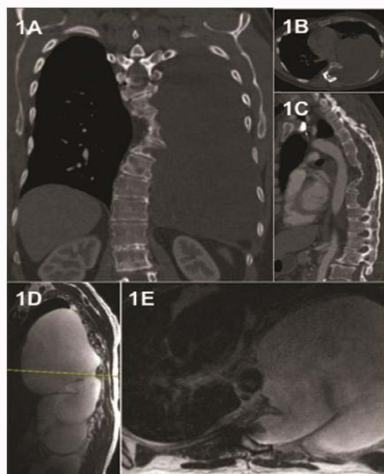


Figure 1: Preoperative CT Chest showing thoracic bony deformities with kyphoscoliosis, vertebral body scalloping on coronal (1A), enlarged thoracic foramen (1B) as well as missing elements of the posterior spinal column on sagittal views (1C). Preoperative MRI Thoracic spine T2 sagittal (1D) and axial (1E) sequences reveals a large fluid-filled cyst arising from the mid-thoracic foramen that is contiguous with the dural sac.

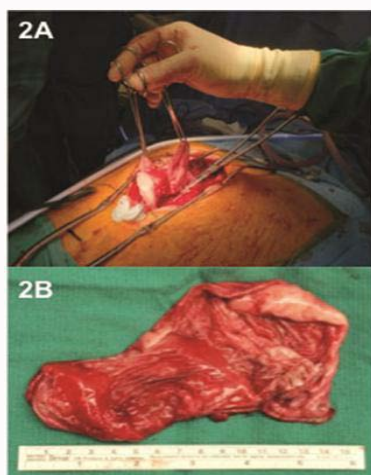


Figure 2: Intraoperative view of the meningocele (2A) from an open, posterior laminectomy and thoracotomy approach. 2B: Partial resection of the bulk of the meningocele from the pleural space measuring 15 cm x 7 cm.

costotransversectomies, and posterior thoracic exploration with Thoracic Surgery for exposure of the meningocele. Single lung ventilation was used during exposure and manipulation of the meningocele. Ligation of the left thoracic nerve roots and additional thoracoscopy allowed visualization of the lateral and anterior edge of the meningocele, which aided in its dissection off the chest wall and visceral pleura. No visceral pleural peel was noted. The base of the meningocele was isolated (Figure 2A) and an endoscopic cutting stapler was used to transect the bulk of the meningocele (Figure 2B). The residual meningocele was plicated at its base and sewn to the lateral border of the vertebral bodies. A pinhole durotomy encountered at the inferior-most thoracic level was covered with a muscle flap. Gelfoam and duraseal were applied over the muscle to complete the repair. Valsalva maneuver demonstrated no visible CSF leak. Two chest tubes were left in the resection cavity and maintained on water seal. For closure, the thoracic paraspinous muscle fascia was dissected to advance the left paraspinous muscle forward onto the



Figure 3: Postoperative MRI Thoracic Spine T2 sequences of the coronal (3A) and sagittal (3B-3D) views showing partial resection of the meningocele.

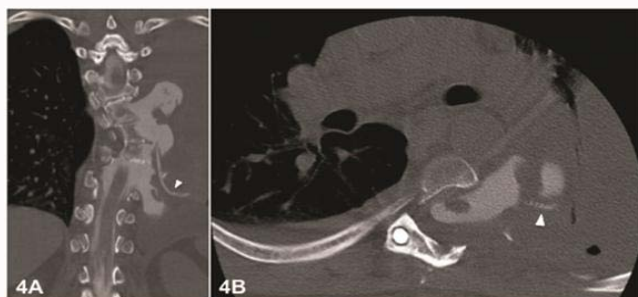


Figure 4: CT myelogram of the thoracic spine showing contrast filling the expected residual meningocele without extravasations into the pleural cavity in the coronal (4A) or axial (4B) views. White arrowheads indicate anastomotic stapler line.

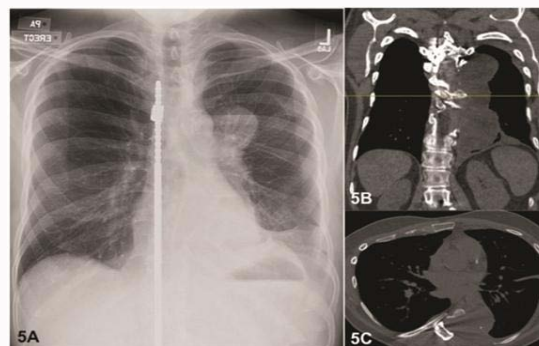


Figure 5: Follow-up CXR AP (5A) at two weeks after discharge and CT Chest (5B,5C) at six weeks show stable size of meningocele.

residual dura and meningocele.

Postoperative course

Postoperatively, the patient had a MRI thoracic spine (Figure 3) which revealed a significant decrease in mass of the meningocele. He was placed on bed rest with lumbar drainage goal of 10 cc/hr. He had daily serial CXRs to monitor for postoperative lung expansion. The patient initially had a small apical pneumothorax that slightly expanded on subsequent imaging. The chest tubes remained on water seal due to concern that suction would strain the surgical scar and potentiate a CSF leak. The chest tubes were removed on postoperative day Day (POD) 5 after output decreased. After one week, the LD was clamped and removed following a CXR that revealed a small pleural effusion without significant compression of the left lung. Unfortunately, subsequent chest imaging revealed increasing size of the pleural effusion. While the patient remained asymptomatic, the presence of the fluid was concerning for a CSF leak. The patient thus underwent a CT myelogram that did not have contrast extravasation into the pleural cavity (Figure 4). A percutaneous small bore chest tube

was placed which drained 1.7 L of fluid. The patient was discharged home after chest tube removal with no neurological or respiratory compromise on POD14.

The patient was seen in follow-up with repeat CXR and CT Chest showing good lung expansion and stable size of the residual meningoceles (Figure 5). He reported some numbness along the left chest wall and some incisional pain but otherwise denied respiratory symptoms.

Discussion

Thoracic meningoceles are rare and even more infrequently require intervention. Management can be variable and dependent on size and the patient's clinical presentation. Smaller and asymptomatic lesions are followed conservatively with imaging. When indicated, surgical options include: 1. drainage of the meningocele via cystoperitoneal shunt or lumboperitoneal shunt [2,10,12]. 2. Resection or reduction via posterior laminectomies or 3. Lateral thorascopic or open thoracotomy approach for resection [2,3,12-17]. Cystoperitoneal or lumboperitoneal shunts, however, can have shunt failure or malpositioning and early recurrence of the cyst [2]. Non-Thoracotomy exposures can avoid chest tubes post-operatively, but provide smaller surgical windows that are restricted to smaller meningoceles and make watertight closures difficult. Thoracotomy exposures offer a wider operative field, but necessitate chest tube drainage and introduce the risk of an intra thoracic CSF fistula [15]. Laminectomy with costotransversectomy can accelerate progression of kyphoscoliosis, leading to neurologic symptoms [3,7]. Concurrent or subsequent posterior fusion must be counseled with the patient. Our patient fortunately had a right-sided Harrington rod and abnormal vertebral bodies with widened foramina corridors, which allowed the authors to approach the meningocele from a prone, posterior thoracotomy approach without concern for subsequent structural instability. This is unique as prior thoracotomies for large meningoceles have all been performed in the lateral decubitus, transthoracic position, and included incisions along the inter space.

Intraoperatively, approach to the meningocele will depend on the dural integrity. Prior authors have cited a CSF leak from the suture site due to the poor dural quality. Options for removal of the meningocele include resection of the entire mass, ligation of the stalk of the meningocele, and puncture and plication of the meningocele to decrease mass burden [2,15]. Lumbar drains are universally used for CSF diversion to reduce tension of repair site. We had the benefit of a wide posterior decompression for access of the lateral and ventral borders of the meningoceles. Resection of the thoracic nerve roots un-tethered the meningocele from the chest wall and allowed the authors to maneuver an endoscopic stapler to transect the bulk of the meningocele. The endoscopic cutting stapler provided a reasonable watertight closure of the meningoceles that was reinforced by plicating the residual meningocele onto itself to conceal the staple line.

Thoracic meningoceles that require surgery tend to be large. As such, a substantial CSF volume can be lost during durotomies that leads to fluid shifts. Similar to Das et al. [2], our patient had roughly 2 L of CSF in his chest cavity that, when released, resulted in hypovolemic shock requiring aggressive resuscitation and temporary packing of the chest cavity. Considerations in patients with large thoracic meningocele may include placement of a central line and arterial line for aggressive volume resuscitation, pressors, and accurate blood pressure readout.

Postoperatively, the patient was placed on antibiotics for one week. While there is inadequate literature to support this, antibiotics were started given the communication between the pleural cavity and CSF. The patient was also on bed rest with a LD and two chest tubes. While there is significant variation across case reports, LDs were typically maintained for a week for CSF diversion away from surgical site. Chest tubes were rarely placed on suction for fear of violating the suture sites and creating a CSF leak or cysto-pleural fistula. The authors removed the chest tubes prior to the LD. Unfortunately; our patient had an early recurrent left pleural effusion, likely related to chronic lung collapse. The decision to not apply negative suction to the chest tube for dura healing also likely contributed to incomplete lung expansion and pleural apposition.

Before treating the pleural effusion, the authors sought to discern any communication between the meningocele and pleural space. T2-weighted sequences on a MRI Thoracic spine demonstrated the residual meningocele but included streak artifact from the Harrington rod. CT Thoracic spine could not discern between fluid in the meningocele and pleural transudate. A CT myelogram ultimately was the best imaging modality to discern small dural defects and was fortunately negative in our case. The pleural effusion was thought to be transudative fluid that accumulated due to inadequate lung expansion. With a CSF leak ruled out, a chest tube was replaced by thoracic surgery, and gentle (-10 cm) suction was applied. Short-interval follow up with serial chest imaging is pivotal in such scenarios where chronic lung compression by the meningocele can delay or hinder lung re-expansion.

Conclusion

Due to its rarity, surgical and medical management of thoracic meningoceles are poorly defined and standardized. Here, we describe some perioperative issues beyond the surgical techniques required for treating a patient with a large and complex thoracic meningocele. While there may not be a singular protocol for treating NF1-associated thoracic meningoceles, knowledge and preparation for intraoperative fluid shifts, varying surgical approaches to minimize postoperative CSF leaks or fistulas, management of lumbar and chest tube drains, as well as postoperative imaging are the key points in successfully addressing such a rare condition.

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