Rupture of Giant Anterior Sacral Meningocele in a Patient with Marfan Syndrome: Diagnosis and Management

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Key words
- Anterior sacral meningocele
- Dural ectasia
- Management
- Marfan syndrome
- Rupture

Abbreviations and Acronyms
ASM: Anterior sacral meningocele
CSF: Cerebrospinal fluid
CT: Computed tomography
MRI: Magnetic resonance imaging

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BACKGROUND: **Marfan syndrome is a genetic disorder that results in the weakening of connective tissues. Dural ectasia has been defined as a feature of Marfan syndrome and is present in up to 92% of patients. Rarely, dural ectasia can erode through the sacrum expanding into an anterior sacral meningocele.**

CASE DESCRIPTION: **Information for this case report was gathered from patient notes and imaging from the patient chart. This is a case of a 46-year-old woman who presented with urinary incontinence, early satiety, and back pain in the setting of a known anterior sacral meningocele. Before operative management, the anterior sacral meningocele ruptured with the patient presenting signs and symptoms of intracranial hypotension. Conservative management did not alleviate the pain. She was ultimately managed with posterior sacroplasty followed by anterior sacral meningocele resection and placement of a lumboperitoneal shunt. The patient did not have reaccumulation of the meningocele or recurrent symptoms at the latest follow-up.**

CONCLUSIONS: **The progression of dural ectasia in Marfan syndrome to an anterior sacral meningocele is uncommon. It is important to identify the characteristics associated with an expanding dural ectasia as this patient's symptoms progressed over time and the meningocele grew large. Given its rarity, there are no guidelines in place regarding size at which repair of an anterior sacral meningocele should occur prophylactically. It is important to review these cases in order continue to learn about progression, management, and outcomes of patients with an anterior sacral meningocele.**

INTRODUCTION

Marfan syndrome is an autosomal dominant disorder affecting 1 in 5000 people.[1] It is caused by a mutation on chromosome 15 in the FBN1 gene encoding fibrillin-1.[1,2] Fibrillin-1 deficiencies have been shown to increase levels of transforming growth factor β (TGF-β), resulting in weakening of elastic fibers and connective tissue.[1] Common manifestations of the disease include thinning of the aortic wall and aortic root aneurysms, elongation of long bones, and hypermobility of joints.[3] In addition, a common feature of Marfan syndrome is dural ectasia, found in up to 92% of patients.[1,4] Dural ectasia is defined as the ballooning or widening of the thecal sac, typically found in the sacral spine region where cerebrospinal fluid (CSF) pressure is greatest.[3,5] The most severe form of dural ectasia is characterized by the presence of an anterior sacral meningocele (ASM) with associated thinning of the sacral cortex and expansion into the retroperitoneum and pelvis.[6] The most common symptoms associated with an ASM include radiculopathy, constipation, urinary obstruction, and postural headaches.[5,7-9] Treatment options include observation and surgical management for larger or symptomatic lesions. In this report, we describe a case of a patient with Marfan syndrome presenting with a spontaneously ruptured giant ASM. To our knowledge, this is the first case report in the neurosurgical literature detailing the diagnosis, management, and imaging of a spontaneously ruptured ASM in a patient with Marfan syndrome.

CASE REPORT

A 46-year-old woman with a past medical history significant for Marfan syndrome and dural ectasia presented to the outpatient spine clinic with a chief complaint of 6 months of urinary incontinence, early satiety, and a notable abdominal mass. She also complained of progressing back pain that began radiating to her groin bilaterally and right leg. This pain was relieved by ambulation. She denied any history of previous intervention of her spine for correction of the dural ectasia. She was diagnosed with the ASM 8 years before presentation and was previously followed by a neurosurgeon at an outside hospital with her last magnetic resonance imaging (MRI) being performed approximately 4 months before her current presentation. On examination, the patient had full strength and intact sensation of her upper and lower extremities. A fluctuant abdominal mass, painless to palpation, was noted in the right lower quadrant (Figure 1). A sagittal T2-weighted fat-suppressed MRI (Figure 2A) and corresponding sagittal computed

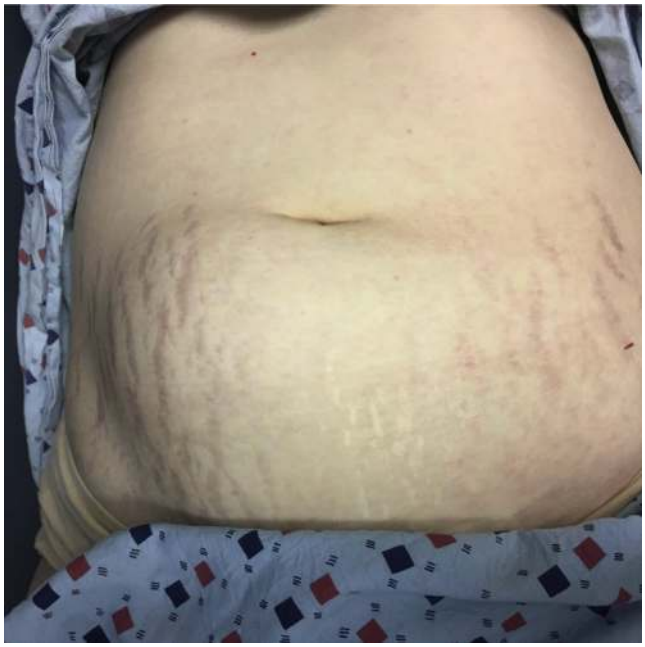

Figure 1. Picture of the patient's abdomen while laying supine revealing a visible palpable fluctuant mass.

tomography (CT) image (Figure 2B) demonstrated dural ectasia with a giant ASM and bladder compression.

Given the size of the ASM and the patient's symptoms, the plan was made to measure CSF opening pressure followed by development of a multidisciplinary approach for repair via posterior and anterior surgeries. However, before initiating this plan, the patient presented to the emergency department with complaints of postural headaches, nausea and vomiting, and decreased intraabdominal fullness. She remained neurologically intact without signs of meningismus. Given her symptoms and loss of abdominal bulge, she was diagnosed with an ASM rupture confirmed on CT (Figure 3). Corresponding MRI (Figure 4) revealed a rupture of the ASM with herniation of bowel and the ruptured ASM sac into the spinal canal. The patient was managed with a 3-stage procedure: 1) sacroplasty with reduction of herniated bowel and titanium mesh reconstruction of the anterior sacral wall (Figure 5), 2) transabdominal resection/repair of the ASM sac, and 3) placement of a lumboperitoneal shunt. The patient was discharged home and at the 1-year follow-up, she was doing well without long-term sequelae.

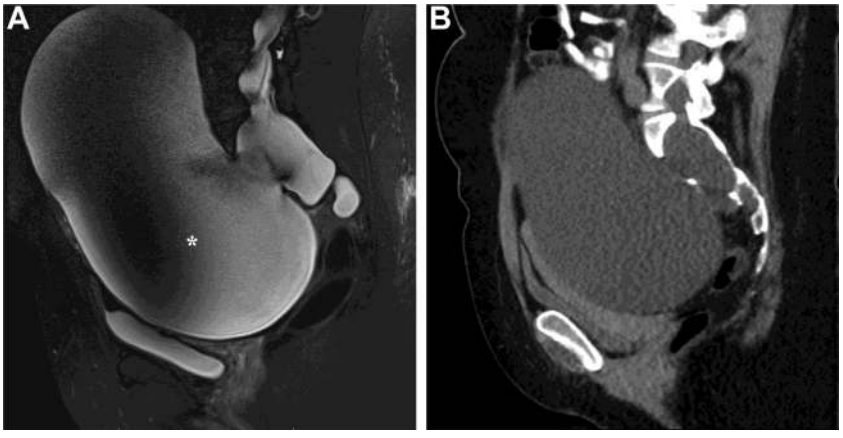

Figure 2. (**A** and **B**) Sagittal T2-weighted fat suppressed magnetic resonance imaging (**A**) demonstrates a giant anterior sacral meningocele (*asterisk*) eroding through the sacrum and compressing the urinary bladder anteroinferiorly. Corresponding sagittal computed tomography image (**B**) demonstrates the anterior sacral bony defect.

DISCUSSION

ASM is a rare condition associated with connective tissue disorders including Marfan syndrome.[1,3,10-13] ASM, most frequently presenting as a presacral mass, is characterized by a herniation of the thecal sac through a bone defect along the anterior margin of the sacrum.[3,8,14] Development of an ASM is caused by normal CSF pulsation on predisposed dural tissue, resulting in progressive growth and eventual erosion of the sacral wall.[5,12,13] CSF pressure is greatest in the caudal regions of the spinal canal, which explains why the sacral region is mostly heavily impacted.[3,5]

ASM is most prevalent in women and can present with a diverse spectrum of clinical findings.[10,15] Although usually asymptomatic in younger individuals, symptoms can begin in early adult life when the presacral mass has grown large enough to exert pressure on surrounding abdominal structures.[10,12,16] Compression of the bladder, rectum, and uterus may cause urinary incontinence, chronic constipation, or irregular menstruation.[7,8,10,17] Postural headaches are present in 10%–15% of patients with an ASM and likely result from intermittent changes in intracranial pressure.[7,17] Rarely, sacral nerve root compression can produce radiating low back and leg pain.[7] In our case, the patient presented with right leg pain, worsening urinary incontinence, and early satiety due to compression of the stomach and surrounding structures.

After a thorough physical examination, imaging tests are essential for defining an ASM. MRI is the preferred imaging modality given its soft tissue contrast

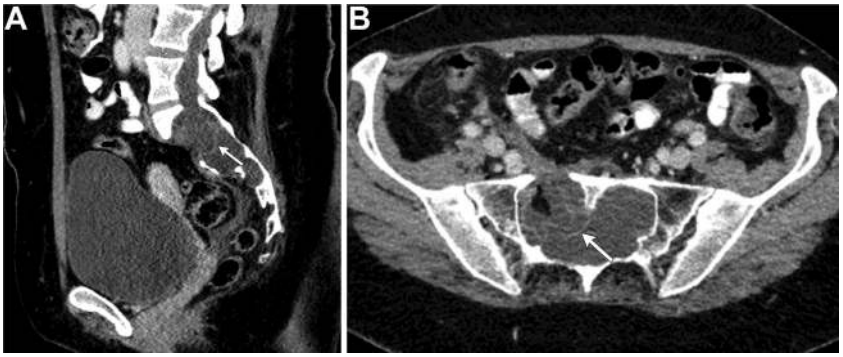

Figure 3. Sagittal (**A**) and axial (**B**) computed tomography images demonstrate interval rupture of the anterior sacral meningocele with herniation of bowel and the ruptured meningocele sac into the sacral canal (*arrows*). In **B,** also note low-attenuation mesenteric fat on the right herniating with the bowel.

superiority and the ability to distinguish nerve roots and details of the ASM structure.[7,15,18] In particular, it is important to be able to define the communication between the ASM and sacral canal. CT evaluation is often used to define sacral anatomy.[3,15] Myelography can be helpful to define communication between nerve roots and the ASM. In this case, the physical examination revealed a firm, nontender palpable abdominal mass (Figure 1). A review of the patient's MRI of the lumbar spine in 2014 and 2016 (Figure 2A) also demonstrated the CSF containing mass, which had enlarged significantly over time. Finally, a review of the CT of the abdomen and pelvis confirmed the existence of a large ASM eroding the anterior sacrum (Figure 2B). Progressive symptoms and these dramatic imaging findings resulted in a more urgent effort to provide the patient with options for definitive treatment.

Treatment options for ASM include observation and surgery depending on the symptoms and size of the lesion. For a small asymptomatic ASM, nonsurgical management is appropriate. This can involve bed rest, focal compression, and abdominal binders.[7] Diversion of CSF fluid via a lumboperitoneal shunt is also a potential treatment option.

The two surgical methods most commonly cited in the literature for resection of an ASM are the posterior midline sacral approach and anterior transabdominal approach.[7,11,17] Introduced by Adson in 1938, the posterior approach involves sacral laminectomy and ligation of the neck of the ASM to close off the communication channel between the ASM and spinal subarachnoid space.[5,17] The posterior approach is often the preferred surgical method, as it allows easy access to the pelvic floor while minimizing potential neurologic or visceral complications.[5,8] On the other hand, the anterior transabdominal approach has a risk of visceral injury and offers poor visualization of the neurologic structures.[8] It has been reported to be associated with a higher complication rate than the posterior approach.[5,8] Nevertheless, the anterior approach is still a safe and regularly employed surgical method, especially in complex cases, large lesions, or those involving adjacent viscera.[7,8,13]

In this case, the patient initially consulted with her neurosurgeon and orthopedic spine surgeon and a posterior-based approach was recommended to reconstitute the anterior thecal sac, as well as buttress the dehiscent sacral bone with a bone graft. The plan included placing a temporary or permanent CSF drainage catheter following the procedure. Less than three weeks following surgical consultation, the patient presented to the emergency department with a rupture of her ASM as demonstrated on CT and MRI (Figures 3 and 4). She noted resolution of her abdominal bulge and improved bladder and bowel continence. However, she also noted development of right buttock pain, postural headache, and nausea. The patient was admitted and underwent sacral laminectomy for ventral sacral reduction of the herniated bowel and

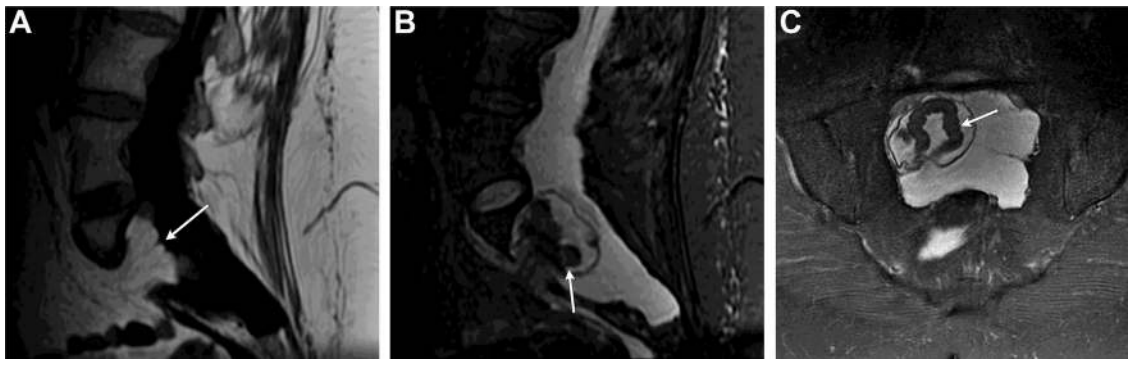

Figure 4. Sagittal T1-weighted image (**A**) demonstrates herniation of high-signal mesenteric fat (*arrow*) into the sacral canal. Corresponding sagittal STIR image (**B**) demonstrates expected low signal of the herniated mesenteric fat (*arrow*) and surrounding low-signal herniated meningocele sac. Coronal T2 fat-suppressed image (**C**) demonstrates the low-signal herniated bowel loop (*arrow*) surrounded by the thin wavy herniated meningocele sac.

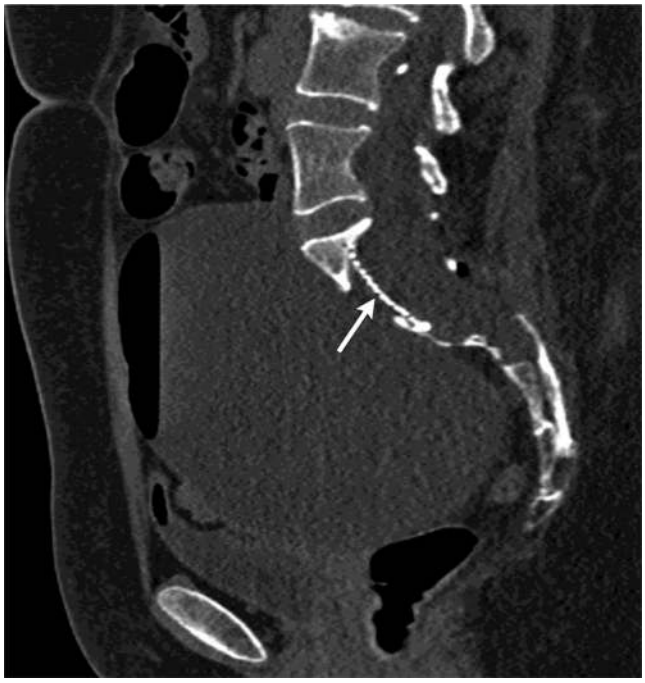

Figure 5. Sagittal computed tomography image demonstrates postoperative sacroplasty with titanium mesh reconstruction (*arrow*). Also note reaccumulation of the anterior sacral meningocele with air fluid level.

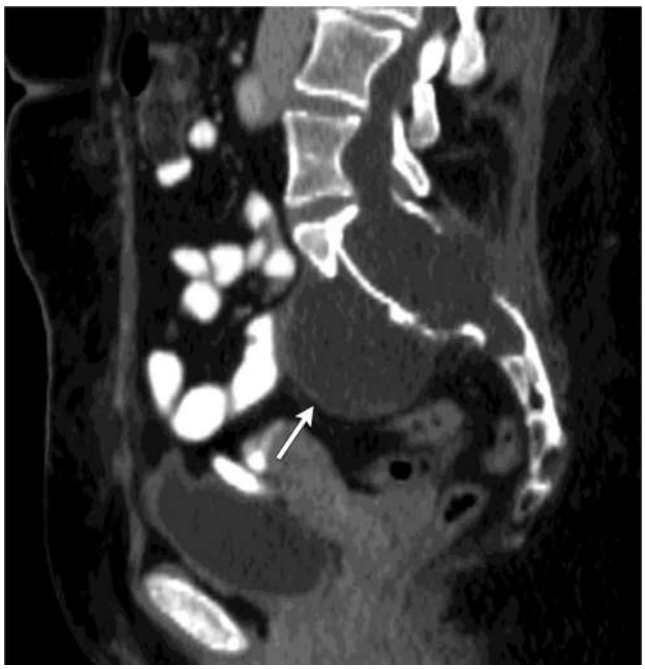

Figure 6. Sagittal computed tomography image demonstrates near-complete resolution of the anterior sacral meningocele (*arrow*) following its resection/repair and placement of lumboperitoneal shunt (not shown).

sacral meningocele sac with titanium mesh graft and AlloDerm dural onlay. A lumbar drain was placed and drained 10 mL/hr of CSF. Following the procedure, the patient was maintained on flat bedrest for three days. A repeat CT showed reaccumulation of the ASM sac, necessitating a second surgery from an anterior approach (Figure 5). A second procedure, which included drainage and resection of the ASM sac and repair of dural tears, was successful. To prevent reaccumulation of the ASM, the patient subsequently had a third and final procedure for placement of a lumboperitoneal shunt (Figure 6). Since completion of her final surgery over 12 months ago, the patient has not had recurrent symptoms.

CONCLUSION

While dural ectasia is present in up to 92% of patients with Marfan syndrome, progression to a giant ASM is rare and there are few guidelines as to standard conservative and operative management. Indications for resection of an ASM include incapacitating headaches and neurologic changes including bowel and bladder changes. If an ASM ruptures, sacroplasty and repair of anterior dural defect can be performed. Given the severity of this condition, it is important to study cases such as this one to understand patient presentation, progression, imaging, management, and outcome.

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