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Giant anterior sacral meningocele associated with hydroureteronephrosis and renal injury: illustrative case

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BACKGROUND Anterior sacral meningocele (ASM) is a defect in the closure of the neural tube. Patients can be asymptomatic or present with genitourinary, neurological, reproductive, or colorectal dysfunction. Magnetic resonance imaging (MRI) is the gold standard test because it can assess communication between the spinal subarachnoid space and the lesion and identify other abnormalities. Surgical correction is the definitive treatment because untreated cases have a mortality rate of more than 30%.

OBSERVATIONS A 24-year-old woman with Marfan syndrome presented with polyuria, recurrent urinary tract infections, and renal injury for 3 months along with a globose abdomen, with a palpable mass in the middle and lower third of the abdomen that was massive on percussion. MRI showed an ASM consisting of two cystic lesions measuring 15.4 × 14.3 × 15.8 and 6.7 × 6.1 × 5.9 cm, respectively, compressing the distal third of the right ureter and causing a hydroureteronephrosis. Drainage and ligature of the cystic lesion were performed. The urinary outcome was excellent, with full recovery after surgery.

LESSONS ASM should be suspected in all abdominal masses with progressive symptoms in the setting of Marfan syndrome. Computed tomography and MRI are important to investigate genitourinary anomalies or other types of dysraphism to guide the best surgical approach.

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KEYWORDS meningocele; dysraphism; spina bifida

Anterior sacral meningocele (ASM) is a rare condition occurring in the presence of a defect in the closure of the neural tube. It is found more frequently in women and consists of uni- or multilocular herniation of the dural sac through a sacral bone defect or an enlarged anterior sacral foramen. The main etiology is congenital, but it can be acquired as a result of dural ectasia, usually associated with specific causes such as Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis types I and II, or anterior sacral trauma.[1–3]

The first description of congenital ASM was published in 1837 by Thomas Bryant, and there are currently just over 250 cases published in the medical literature. The clinical presentation can vary from asymptomatic to the presence of nonspecific symptoms and signs such as constipation, dysmenorrhea, dyspareunia, urinary retention, urinary incontinence, dysuria, polyuria, radiculopathy, and/or paresthesia related to genitourinary, neurological, reproductive, or colorectal dysfunction due to mass effect on the abdominal viscera. There is also an association between this dysraphism and congenital abnormalities such as anorectal malformations, sacrococcygeal teratoma, uterine duplication, lipoma, and dermoid and epidermoid cysts.[1–3]

Magnetic resonance imaging (MRI) is the gold standard test for diagnosing this defect because it is able to assess the communication between the pedicle and the lesion, spinal cord anchorage, and the presence of associated neoplasms. Computed tomography (CT) and lumbosacral radiography can be complementary for better visualization and identification of bone lesions.[1,2]

Because of the lack of spontaneous regression of the sacral meningocele, surgery is the treatment of choice for therapeutic resolution. Surgical approaches can be performed laparoscopically or endoscopically or from a posterior sacral access. However, during childhood, the lesion may be conservatively monitored in asymptomatic cases

ABBREVIATIONS ASM = anterior sacral meningocele; CSF = cerebrospinal fluid; CT = computed tomography; MRI = magnetic resonance imaging.
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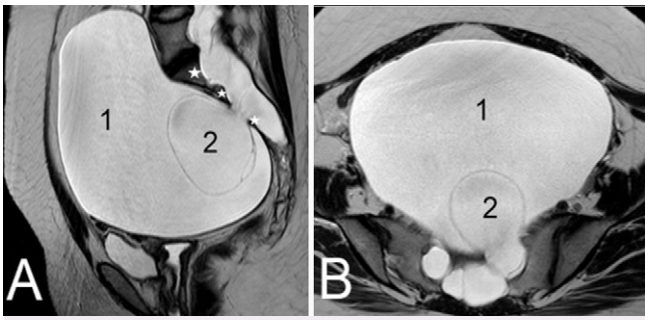

FIG. 1. T2-weighted MRI of the pelvis showing anterior sacral meningocele with two cystic cavities (*1* and *2*), with connection to the spinal space S2-S3 (**A,** 2nd and 3rd *white stars*).

because of the possibility of iatrogenesis leading to urinary or neurological disorders. In untreated cases or those with delayed diagnosis, mortality is more than 30% when associated with infection, especially meningitis. The goal of treatment is to eliminate communication between the meningocele and the spinal subarachnoid space.[1,2,4]

Illustrative Case

A 24-year-old woman diagnosed with Marfan's syndrome presented to our institution with polyuria, recurrent urinary tract infections, and increased nitrogenous waste (creatinine 2.1 mg/dL, urea 82 mg/dL, and creatinine clearance 42.39 mL/min) for 3 months with progressive worsening of urinary symptoms. During diagnostic investigation, an abdominal cystic lesion extending from the intraspinal canal at the sacral level was found, and the patient was referred for neurosurgical evaluation and management.

On examination, the patient had a globose and flaccid abdomen that was painless on palpation and had a palpable and massive mass in the middle and lower thirds of the abdomen. She has had bilateral amaurosis for 7 years as a result of optical impairment due to Marfan's syndrome and has no motor or sensory deficits.

Pelvic T2-weighted MRI showed an abdominal cystic lesion measuring 15.4 $\times$ 14.3 $\times$ 15.8 cm, with another one inside measuring 6.7 $\times$ 6.1 $\times$ 5.9 cm, connected with the spinal cord subarachnoid space at level S2-S3 with a 1.9 $\times$ 1.5 cm pedicle, suggestive of anterior sacral meningocele (Fig. 1). The lesion had a compressive effect on the distal third of the right ureter, promoting significant dilation and tortuosity of the right ureter, classified as grade V vesicoureteral reflux, and a pelvic and calyceal dilatation with parenchymal thinning of the right kidney, classified as grade IV hydronephrosis based on the Society for Fetal Urology grading system (Fig. 2).[1,2]

Surgical Technique

Before surgical procedure, the patient underwent a lumbar puncture, receiving 2 mL of fluorescein for better intraoperative visualization of the spinal defect. The surgical approach used the Cherney incision, consisting of an incision 2 to 3 cm above the pubic symphysis that gave access to the retropubic space, with identification of an extensive cystic lesion with compressive effect on the uterus, the bladder, and the intestines after opening the parietal peritoneum. The lesion was carefully exposed with visualization of its posterior aspects (Fig. 3). An incision of 1 cm was made on the most prominent area of the mass, and cerebrospinal fluid (CSF) was drained with the use of an aspirator. The fluid was collected and isolated in the aspiration recipient; its volume at the end of the procedure was approximately 1,500 mL. A second cystic lesion with fluorescein-stained content was found inside the bigger one, with a pedicle directed toward the sacral bone defect (Fig. 4). Ligature of the mass pedicle was performed in four steps: (1) exposition of redundant layers of the first and second cysts walls, (2) approximation of the edges of the lesion with single stitches using 4.0 Prolene, (3) strangulation of the pedicle using 4.0 Prolene, and (4) removal of the second cyst after the closure of its connection with the subarachnoid space on the previous step. Because the ligature was so tight and there was no CSF leak, we did not need any kind of dural substitute.

At the end of the pedicle closure, we performed a Valsalva maneuver that did not produce any signs of CSF leak, making it unnecessary to use dural sealant. During hemostasis and synthesis of the surgical site, no drain was placed and there was no need to maintain an external lumbar drain because the pedicle was closed hermetically.

The procedure ended without any complications, and the patient remained in the supine position for 24 hours. She was allowed to sit and walk on the following day and was discharged from the hospital 48 hours after the surgery. She returned at 2 weeks, 1 month, 3 months, and 8 months after myelocele correction, with no complications in the postoperative period and with improved renal function (creatinine 0.9 mg/dL, urea 41 mg/dL, and creatinine clearance

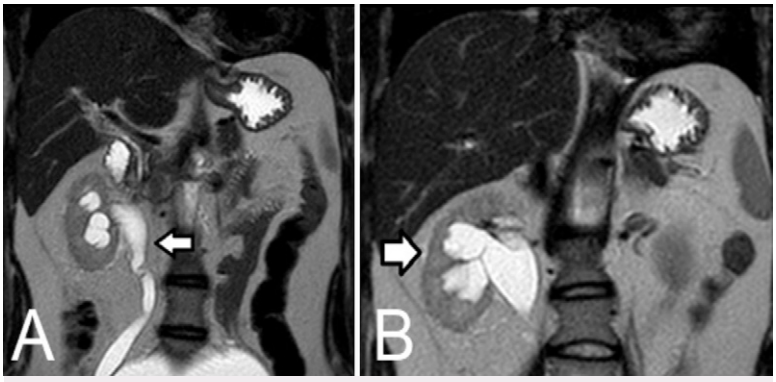

FIG. 2. Abdominal T2-weighted MRI demonstrating grade V vesicoureteral reflux with hydroureter (*arrows*).

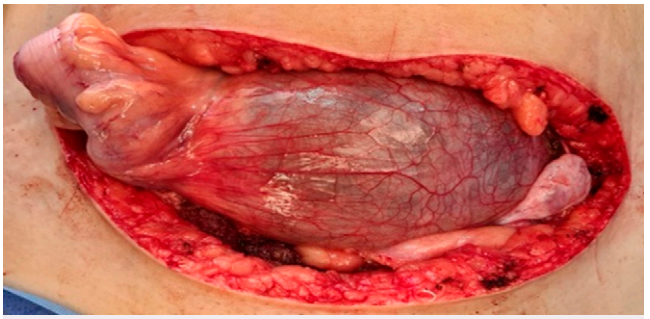

FIG. 3. First cystic lesion seen right after the peritoneum incision.

98.9 mL/min) and no new episodes of lower urinary tract infection since the first month after surgery. A new MRI of the abdomen and pelvis, performed 1 month after the procedure, showed evidence of resolution of meningocele, ureteral compression, and hydronephrosis (Figs. 5 and 6).

Discussion

ASM is a rare form of spinal dysraphism defined as a herniation of the dural sac due to a defect in the anterior portion of the sacrum.[5] When compared to posterior and lateral meningoceles, ASM has a strictly lower incidence, with little more than 250 scientific papers written on this pathology since its first description in 1837.[6]

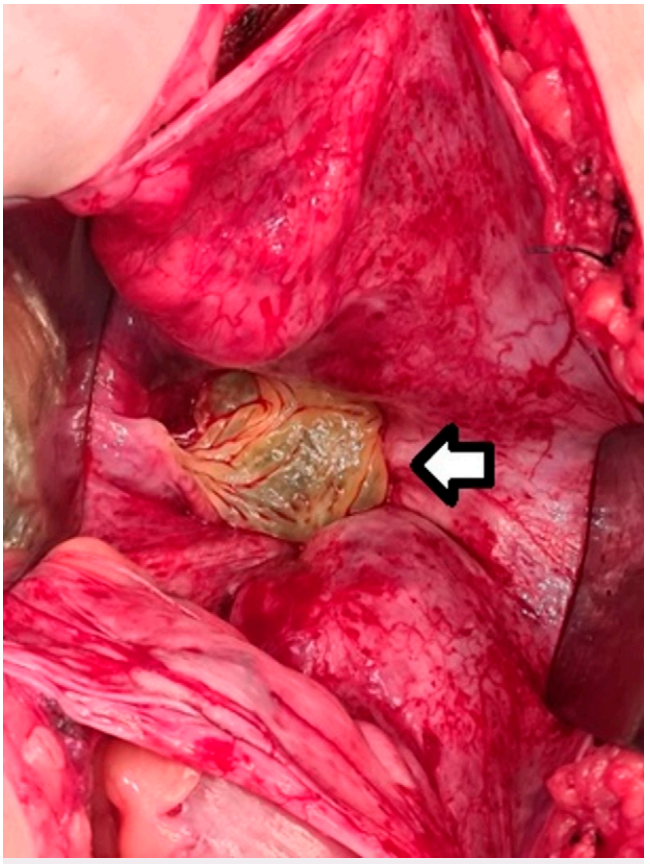

FIG. 4. Second fluorescein-stained cystic lesion visualized after drainage of the first lesion (*arrow*).

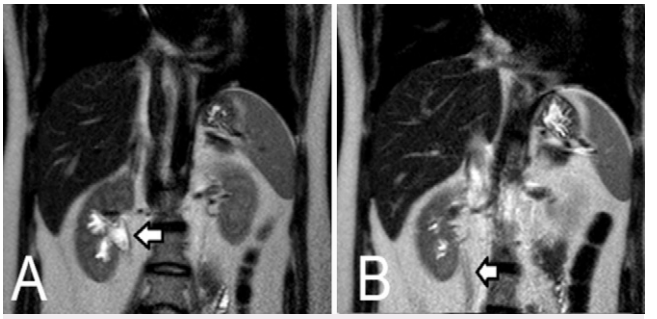

FIG. 5. Abdominal T2-weighted MRI showing resolution of hydronephrosis at the level of the renal pelvis (**A,** *arrow*) and right ureter (**B,** *arrow*).

Unlike posterior meningoceles, which are identified and treated in early childhood, anterior dural closure defects are more frequent in women of childbearing age, which is consistent with our report.[7] They are usually found incidentally or as cystic masses during pelvic or rectal examination for unrelated causes.[5]

The most important precursor factor is dural ectasia. This condition is most frequently found in the caudal neural canal as a result of greater subarachnoid pressure found in this location.[8] The physiological pulsation of CSF is believed to promote expansion of the compromised dural sac, generating lumbosacral vertebral erosion, arachnoid cyst, or anterior meningocele.[8] As the cystic cavity expands and accumulates CSF throughout life, it starts to exert a mass effect on abdominal structures, which can generate symptoms both by compression and by local irritation.[1,2] Some pathologies are associated with lower dural sac resistance, such as Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis types I and II, and anterior sacral trauma. Of these, the most frequently associated is Marfan syndrome, as in the case of our patient.[1,2,5,8]

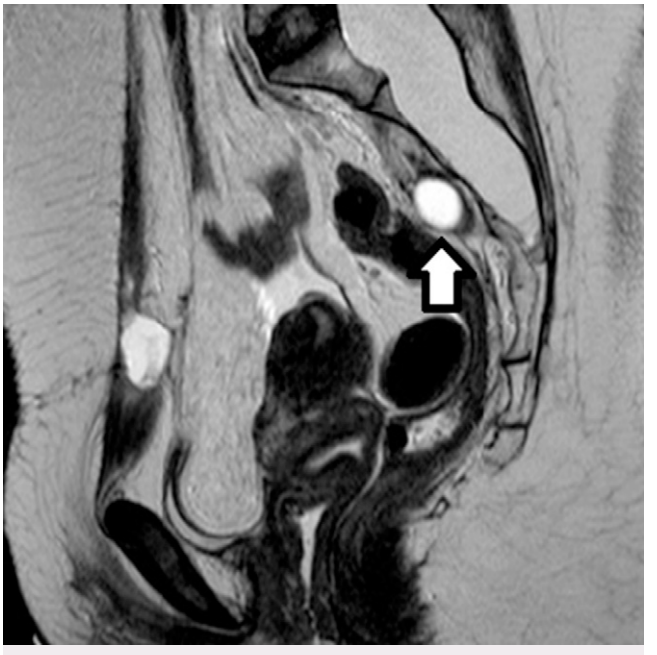

FIG. 6. Abdominal T2-weighted MRI showing resolution of the meningocele with a small residual lesion not connected to the subarachnoid space (*arrow*).

Marfan syndrome is a hereditary disease whose manifestations occur in the connective tissue as a result of an alteration in the secretion of fibrillin-1, a component of the microfilaments of elastic fibers. The main alterations are lens dislocation, aortic dissection and multivalvular insufficiency, arachnodactyly, scoliosis, and dural ectasia.[9] Pyeritz et al. prospectively evaluated 57 patients with Marfan syndrome and observed that 67% of them had dural ectasia, most being asymptomatic and without progression to meningocele.[8,10]

Observations

Most cases progress asymptomatically or through nonspecific symptoms due to compression of abdominal structures, such as constipation, urinary or reproductive disorders, or headache.[4,8] Neurological complications are rare, but these lesions can cause sepsis, meningitis, and infertility in addition to sphincter disorders. ASM is frequently associated with other anomalies such as stenosis or anal atresia, sacrococcygeal teratomas, malformations of the renal system, and dermoid tumors as well as Currarino syndrome.[4,5] Only two cases in the medical literature describe the presence of hydronephrosis associated with this dysraphism and, of these cases, only one reports the onset of renal failure, as demonstrated in our study.[9,11]

Several diagnostic tests can be used both for diagnosis and evaluation of functional impairment, such as barium enema, excretory urography, myelography, CT, and MRI.[4] MRI is the gold standard test for the diagnosis of this defect because it is successful in assessing communication between the pedicle and the lesion, the spinal anchorage, and the presence of associated neoplasms. CT and lumbosacral radiography can be complementary for better visualization and identification of bone lesions.[1]

There is no grading system for anterior meningocele size, but we classify ours as a giant meningocele because it is the second biggest lesion when compared to other papers that call their masses giants.[3–6] As far as we know, our study is the first to identify two cystic lesions in the same patient, one inside the other, and the first to use and describe the use of fluorescein. We used fluorescein to search for a connection between the two cysts, observing that there was none and that their contents did not mix.

During surgery, we observed that the wall of the bigger mass had a fibrotic aspect, similar to the dura mater, and that the smaller wall was translucent like a thickened arachnoid layer. Both walls were sent for analysis, confirming that the outer cyst was made of dura mater and the inner one of arachnoid. Because the arachnoid is permeable to CSF and, to some extent, produces its own CSF, we do not know if the liquid accumulation between the two layers was due to extravasation from the inner cyst or produced from its wall. However, the absence of fluorescein in the first mass favors the hypothesis that the second wall produced the contents of the bigger cyst.

The symptoms of anterior meningocele do not resolve spontaneously and may progressively worsen, depending on the expansion of the lesion and its compressive effect, as well as increase the risk of infertility, sepsis, and meningitis.[4,12] According Fitzpatrick et al., mortality in patients without surgical treatment can reach 30%.[3]

The objective of surgical treatment is to obliterate communication between the meningocele and the sacral subarachnoid space; a hermetic closure of the meningocele pedicle is recommended. Surgical planning should include research for thickening of the filum terminale, size of the lesion's pedicle, and presence of presacral masses or other associated malformations before and during the procedure.[4]

The first surgical attempts consisted of draining the cyst lesion through the transvaginal or transrectal approach, but most cases resulted in meningitis, greatly increasing mortality and morbidity. Other surgical approaches were developed, but so far, there is no consensus regarding which would be the best strategy for the treatment of ASM. Various alternatives include the anterior approach, such as open or endoscopic access (spine endoscopy or laparoscopy), and the posterior approach, such as through open sacral laminectomy or via endoscopy.[13]

Sacral laminectomy ensures better exposure of the cyst pedicle and sacral nerve roots and exposes possible associated malformations; however, it is more invasive, requires an extended surgical time, does not consider associated pelvic masses, and enlarges the existing bone defect. Meanwhile, the endoscopic approach, first described in 1922,[14] is minimally invasive and equally effective for defects with ostia smaller than 1 cm and uses less exposure, but on the other hand it has a lower success rate in lesions with larger pedicles.[13]

Anterior open access is recommended in meningoceles with wide ostia (larger than 1 cm) or if there is an associated pelvic mass because of greater exposure of the surgical field. Another indication is the absence of associated spinal or anorectal malformations. An advantage of the anterior approach is the access safety because once the cyst is drained, the surgical corridor follows inside its remaining cavity, reducing the manipulation of abdominal structures.[13] In our case, we used this surgical access because of the size of the lesion; the absence of posterior medullary, bone, and anorectal malformations; and the size of the meningocele pedicle, which was more than 1 cm.

Laparoscopic and spinal endoscopy options in anterior approaches are also effective for the synthesis of the meningocele pedicle, but like the posterior approach, they are limited by lower exposure of the surgical field and adjacent structures and the lower success rate in cysts with larger ostia.[6,7,13,15]

Lessons

Our work describes a rare case of a giant anterior sacral meningocele containing two cystic lesions in a 24-year-old woman with Marfan syndrome who presented with polyuria, recurrent urinary tract infections, and renal dysfunction for 3 months as a result of hydroureteronephrosis due to stenosis of the right ureter by the dysraphic mass. This case reinforces the long-term risks of patients with Marfan syndrome, especially women, of developing recurrent infections and dysfunction of abdominal organs, such as the bladder and the kidney in our patient. The anterior abdominal approach is a safe and resolutive way to interrupt the connection between the spinal dural sac and the dysraphism, thus ensuring complete drainage and removal of the meningocele.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Loiola, Guedes. Acquisition of data: Henriques, Moreira, Gregório. Analysis and interpretation of data: Henriques, Loiola, Guedes. Drafting the article: Henriques, Loiola. Critically revising the article: Henriques, Loiola, Guedes. Reviewed submitted version of manuscript: Loiola, Moreira, Gregório, Schmidt. Statistical analysis: Henriques, Schmidt. Administrative/technical/material support: Loiola. Study supervision: Moreira, Gregório, Vasconcelos, Guedes.

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