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CASE REPORT

Anterior sacral meningocele presenting as intracystic bleeding

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Abstract
Purpose To report a case of anterior sacral meningocele with intralesional bleeding secondary to sacrococcygeal trauma. Likewise, there is a discussion about the physiopathology and the surgical approach to these types of lesions.
Methods A 43-year-old man diagnosed with Marfan syndrome suffered sacrococcygeal trauma. He was admitted to the emergency room due to symptoms of headache, nausea, and lower limb subjective weakness. CT and MRI showed a large retroperitoneal mass with hemorrhagic content close to the sacrum. Likewise, the MRI showed an image compatible with subarachnoid hemorrhage in the thoracic spinal area, cerebral convexity, and the basal cisterns. The patient went into surgery for an anterior abdominal approach in the midline to reduce the content of the lesion, and subsequently, in the same act, a posterior approach was done with an S1–S2 laminectomy and obliteration of the pedicle. Postoperative MRI 5 months later showed resolution of the ASM.
Results Anterior sacral meningocele is characterized by herniation of the dura mater and the arachnoid mater outside the spinal canal through a defect of the sacrum. We add the risk of bleeding after trauma—never seen in the literature—as one of the possible inherent complications of this lesion.
Conclusions This report highlights a complication never seen in the literature of a relatively rare condition. In our case, the combined approach was effective for both clinical control and lesion regression.

Keywords Anterior sacral meningocele · Marfan syndrome · Trauma · Hemorrhage · Surgery

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Introduction

ASM is characterized by a herniation of the dural sac through a bone abnormality on the anterior surface of the sacrum. One of the multiple underlying factors may be the dural ectasia [23, 28] related to connective tissue diseases, such as Marfan syndrome. We report a case of ASM with intralesional bleeding secondary to sacrococcygeal trauma.

Case report

History and examination

A 43-year-old male diagnosed with Marfan syndrome anticoagulated for aortic mechanical valve suffered sacrococcygeal trauma. Two days later, he was admitted to the emergency room due to 24-h symptoms of headache, nausea and lower limb subjective weakness as the coccygeal and hypogastric pain escalated. Additionally, the patient presented with a sensation of having a full stomach and difficulty urinating.

Exploration did not show loss of sensation or motor skills. Reflexes were normal and symmetric. Positive Kerning's sign.

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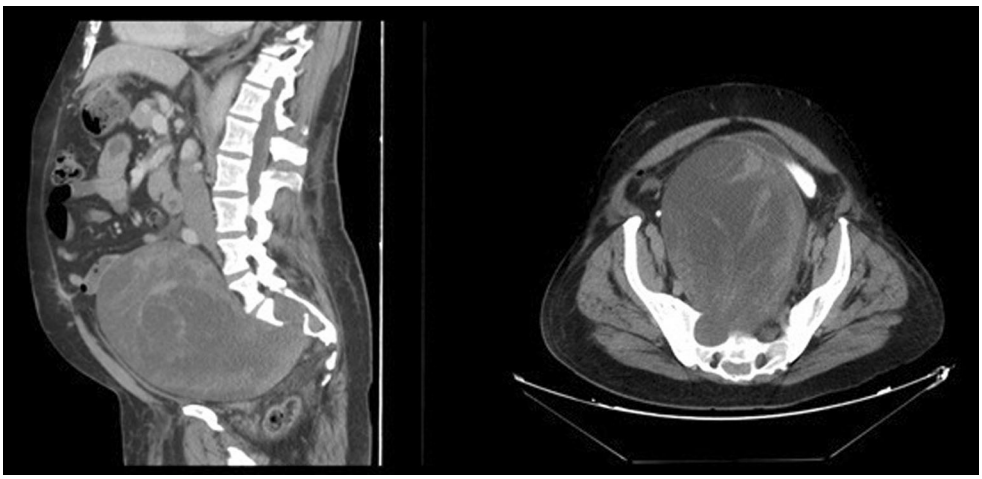

Fig. 1 Preoperative lumbar spine CT showing a large retroperitoneal mass. The mass presents hemorrhagic content and it seems to communicate with the spinal canal through an enlarged right S1–S2 foramen with no content inside

A bladder catheter was inserted for urine retention and hematuria. CT and MRI showed a large retroperitoneal mass causing displacement and compression of the abdominal structures (Figs. 1, 2). Furthermore, the mass presented hemorrhagic content and was extremely close to the sacrum (Fig. 2a, b). Likewise, the MRI showed an image compatible with subarachnoid hemorrhage (SAH) in the thoracic spinal area, cerebral convexity and basal cisterns, which shows the communication between the cyst and the spinal canal (Fig. 2c, d). Lab tests showed anemia and acute kidney failure secondary to hydronephrosis caused by extrinsic compression of the urinary tract, resolved with double J stent.

Surgical procedure

Given the symptoms resulting from the cyst's mass effect the patient went into surgery for an anterior abdominal approach in the midline to reduce the content of the lesion. 1500 cc of dark liquid was drained (Fig. 3a) and a defect at the bottom of the lesion was detected. No neural elements were found entering the ASM. Since an anterior ligation was not possible due to multiple adhesions, the accessible part of the cyst was resected and a plication was performed at the base (Fig. 3b). Subsequently, in the same act, a posterior approach was done with S1–S2 laminectomy (Fig. 3c) and obliteration of the pedicle with autologous fat, fibrin glue and a fibrinogen-thrombin graft.

Postoperative course

Postoperative MRI 5 months later showed resolution of the ASM (Fig. 4). He developed initial post-surgical urinary and bowel incontinence and complained of perianal numbness which improved within 6 months.

Discussion

ASM is a herniation of the dura mater and arachnoid mater outside the spinal canal through a defect either in the anterior sacral wall, or anterolaterally, through an enlarged intervertebral foramen, resulting in a cystic structure generally located at the presacral extraperitoneal space, filled with cerebrospinal fluid and, occasionally, neural elements and/or benign dysplastic tissues or tumors, and is continuous with the spinal subarachnoid space [1–3, 7, 9, 11, 14, 15, 18, 19, 21–23, 30, 32]. ASM may be congenital or acquired and in most cases the defect is a smooth oval structure with well-defined edges [15].

Fewer than 350 cases have been reported in the literature [1, 2, 6, 9, 11, 15, 16, 21, 26, 31]. Equally, it has been related to Currarino syndrome [19, 30] and the history of zero syndrome cases in multiple members of the same family. The meningeal sac is generally formed by two layers, an outer dural membrane and an inner arachnoid membrane [7, 24]. In acquired lesions, the proposed pathogenic mechanism consists of a pulsatile stress that expands a predisposed dural tissue, which progressively grows resulting in bone erosion due to pressure fluctuations of the cerebrospinal fluid (CSF) [6, 12, 18, 19, 24, 27, 30]. The highest pressures occur on the lower areas of the spinal canal, explaining the largest impact on the sacral region [24]. It has also been described as the result of elevation in CSF pressure caused by spontaneous SAH [29] leading to an enlargement of a preexisting dural ectasia, characteristic of Marfan syndrome. Such was the case reported by Strand in 1971, in which the compatible clinical presentation of subarachnoid hemorrhage occurred 2 months before meningocele diagnosis. However, in our case, the traumatic phenomenon precipitated secondary bleeding that does not seem to be the cause of the mass, but the way this

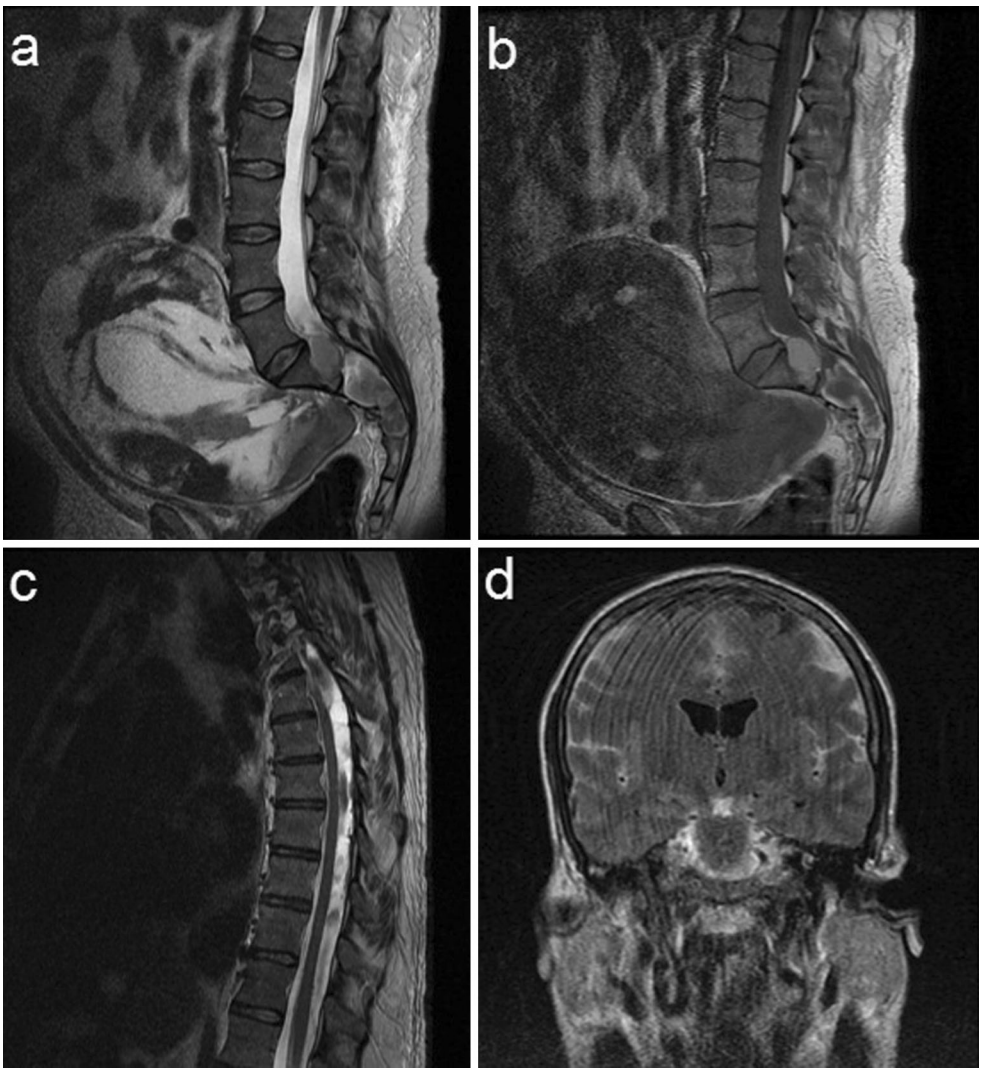

Fig. 2 Different MRI cuts show a presacral mass, measuring 20 × 16 × 14 cm, with apparently no elements inside the cyst, hyperintense in T2 (**a**) and hypointense in T1 (**b**) with images of intralesional bleeding and clear communication with the spinal canal in the sacral region contiguous to the S1 right foramen. It also shows an image compatible with SAH at the thoracic spinal area (**c**), cerebral convexity and basal cisterns (**d**)

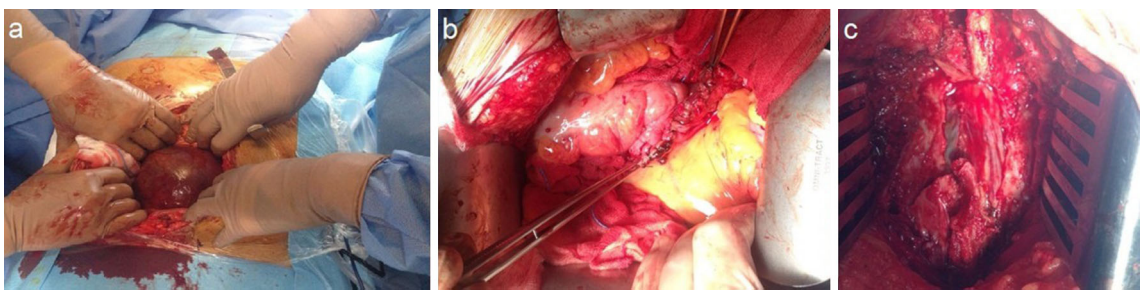

Fig. 3 Intraoperative image through the anterior abdominal approach showing the ASM dome (**a**). After draining it, we proceeded with sac resection and plicature at its base. The image clearly shows peritoneal–retroperitoneal adherences (**b**). Posterior transsacral approach with S1–S2 laminectomy to proceed with pedicle isolation (**c**)

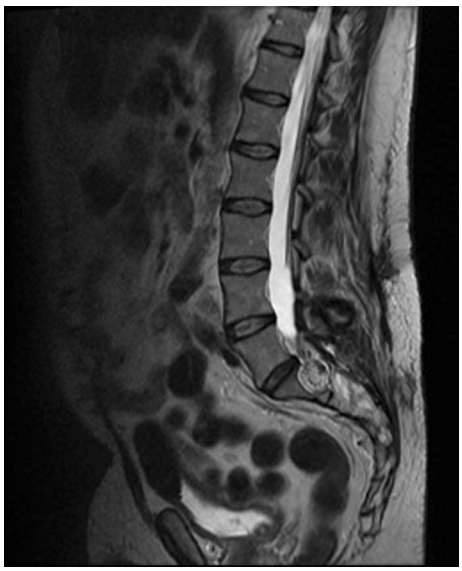

Fig. 4 Postoperative MRI 5 months after surgery showed practical resolution of the ASM

preexisting lesion manifested itself. Similarly, traumas leading to dura mater rupture will result in a pseudomeningocele, filling large spaces [28] with the same pulsating erosion mechanism [5, 30].

The associated symptoms are generally rare and mild [10], which is usually the cause of a late diagnosis [19]. The size of the cyst increases as the patient gets older [8, 18, 30]. The clinical manifestations are commonly constipation, urinary tract disorders and abdominal distension [1, 2, 7, 11, 13, 15, 16, 18, 19, 21, 23, 24, 28, 30–32]. Postural headache due to intracranial pressure variations can result either from hydrostatic pressure directly on the cyst as a result of the Valsalva manoeuver transmitted in retrograde to the intracranial region, or from hypotension due to positional changes [1, 28, 32]. These lesions do not regress spontaneously, and the risk of serious complications, meningitis included [7, 8, 13, 16, 19, 22, 23, 30] leads to consider surgical intervention from the beginning [1, 21].

The goals of surgery should be decompression of the pelvis through drainage and/or excision of the cyst, obliteration of the communication between the cyst and the subarachnoid space and resection of associated tumors, and detethering of the spinal cord, when necessary [19, 23, 25]. The main principles of surgical procedure are capsule delimitation, drainage of its content, reduction of any neural elements within the dura, and primary closure of the dural defect [28]. The CSF lumboperitoneal shunt may be used as an adjuvant to the treatment in case the primary closure fails [30].

Transvaginal and transrectal needle aspirations are contraindicated because of high secondary morbidity and mortality, especially infections [2, 6, 7, 9, 16, 19, 22, 30–32].

The posterior transsacral approach, introduced by Adson in 1938, exposes communication with the meningocele. It involves aspirating its content through the pedicle and closing the communication with primary ligation or obliteration with a graft [2, 3, 7, 8, 10, 11, 13, 14, 18–20, 29–32]. After the fluid has been aspirated, the sac may be left in the pelvis; it is considered unnecessary to resect the meningocele since, after its isolation [10, 13, 18, 30], it will be absorbed gradually [13]. This approach is the most widely used by neurosurgeons and it has been traditionally considered as low-risk and with fewer complications. It will always be the first choice when possible, as in the case of non-associated tumors, because it allows better access to the pedicle and protection to the sacral roots, lower risk of infection and morbidity, and it allows for detethering when needed [2, 6–9, 11, 13, 16–19, 22, 31].

Transabdominal excision with obliteration of the communication [2, 6, 9, 11, 13, 16, 19, 29, 30, 32], developed in 1918, allows direct exposure of the lesion [30]. This approach is very helpful with voluminous mass lesions [19]. Nevertheless, given that meningocele dissection and access to the pedicle are very complex [11, 19], this approach does not allow for a proper manipulation of the nervous structures [19]. In cases where the neck and ostium of the cyst are large, or where they are adhered to the rectum and cannot be dissected, ligation and transdural suture are extremely difficult [11]. This approach is considered an option in cases with associated tumors [16, 31]. Moreover, it is especially useful in cases of large lesions with a significant communication defect [30, 31] because a huge ASM may be challenging in the prone position; therefore, the patient may need cyst reduction through an anterior approach before to the transsacral posterior approach [31]. It is not recommended in the presence of neural elements in the cyst [31]. Due to the close adherence of the sac to the posterior rectum wall, an extensive resection of the dural capsule should be avoided; instead, it is best to perform a limited sac removal [13, 25].

In the sagittal posterior approach [7, 13, 19, 25], the midline incision reduces the risk of neurological complications [7, 30], and the striated muscles can be identified and divided without interrupting their fibers through electric stimulation [19, 31]. It is not recommended in cases of high sacral implantation of the pedicle [31].

The use of endoscopy [4, 13, 19, 25, 28, 30] has been recommended as an alternative, especially for non-obese patients with small fistulous connection.

Several months of follow-up are required to verify the reduction of the cyst, and if it is still present in early post surgery recovery it does not necessarily mean a new surgery is needed [31].

We add the risk of bleeding after trauma—never seen in literature—as one of the possible inherent complications of this lesion. This is most seen in anticoagulated patients, which is especially common in those with Marfan syndrome because they frequently carry mechanical heart valves. Hence, this will support the option of surgical treatment even with asymptomatic patients. The combined approach was effective for both clinical control and lesion regression.

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

The patient has consented to submission of this case report to the journal.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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