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Multidisciplinary team approach in management of anterior spinal dysraphism with unusual presentation: case series and surgical approach

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Abstract

Objectives To report our experience with three cases of anterior lumbosacral meningocele with our multidisciplinary team plan and perioperative management and surgical approach for each case.

Background Anterior meningocele is a rare congenital form of spinal dysraphism that involves protrusion of the theca anteriorly into the retroperitoneal and presacral space through an anterior defect.

Method Three cases with anterior meningocele at different spinal levels (lumbar and sacral) were operated at Neurosurgery Department at Assiut University hospital from June to November 2020 with multidisciplinary management plans that were tailored separately for each case.

Conclusion Anterior meningocele is a rare form of spinal dysraphism and a challenging congenital anomaly that requires proper diagnosis and selection of surgical approach. Our case series outlines the importance of multidisciplinary team approach for tailoring perioperative management and surgical approach with intraoperative important steps.

Keywords Amenorrhea, Marfan syndrome, Multidisciplinary team, Anterior spinal meningocele

Introduction

Anterior spinal meningocele (ASM) is a rare congenital form of spinal dysraphism that involves protrusion of the theca anteriorly into the retroperitoneal and presacral space through an anterior defect [1]. They are typically associated with neurofibromatosis type I but can occur in isolation [2].

Anterior meningocele can be asymptomatic or may manifest clinically. Symptoms are caused by its' mass effect on surrounding pelvic structures, which causes chronic constipation, urine problems and gynecological problems among other things. The combination of obstructive renal failure and hydronephrosis caused by large ASMs is uncommon [3]. The majority of authors selected posterior strategies in their surgical approach due to surgeon's familiarity with this approach, with only a few examples of anterior procedures described. However, due to shortage of studies, there are no precise

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guidelines on how to choose an approach for a patient [3].

Our aim is to provide a surgical plan for each type of anterior meningocele, perioperative management and multidisciplinary team cooperation to maximize patient safety and decrease recurrence and morbidity.

Methods

Three cases with anterior spinal meningocele at different spinal levels (lumbar and sacral) were operated at Neurosurgery Department at Assiut University Hospital through the period from June to November 2020.

All patients underwent a complete admission history, physical examination and standard screening laboratory work. Complete neurological examination was done for all patients. Collecting data during admission included age, sex and complaint.

All patients were admitted to the hospital before the procedure and operated within 48 h of admission. All patients were operated under general anesthesia.

Multidisciplinary management plans were tailored separately for each case. All patients were operated through anterior abdominal approach.

All patients were followed up clinically and radiologically immediate postoperative and 6 months postoperative.

The primary outcome was evaluated as operative complications such as neurological morbidity (irrespective of duration), the presence of postprocedure hemorrhage (subdural, extradural, cerebral and subarachnoid hemorrhage), CSF leak or infection (wound infection, meningitis and pneumonia). The secondary outcome was evaluated by modified Rankin scale (MRS) at discharge.

Postoperative MRI was done within 6 months.

Results

The study had three cases with anterior spinal meningocele two at lumbar level and one case at sacral level. The three cases were females. Two of our patients (66%) complained of abdominal swelling, and one patient 33% complained from menstrual irregularities. The case which was complaining of menstrual irregularities was misdiagnosed as an ovarian cyst and was operated by a general, a surgeon colleague and the patient suffered from foot drop postoperatively, which did not improve after our surgery, but her menses became regular. None of our cases had sensory affection, but 33% had frequency of urine which improved postoperatively. All cases were operated via anterior abdominal approach with multidisciplinary team plan tailored individually for each case (Table 1)(Figs. 1, 2, 3).

Discussion

Anterior spinal dysraphism is a rare pathology that had been discussed few times in the literature without a definite management protocol. The ideal aim of the surgery is complete excision of the sac with direct repair without any intraoperative or postoperative complication like recurrence or CSF leak.

Marfan syndrome (MFS) is a spectrum of disorders caused by a heritable genetic defect of connective tissue that has an autosomal dominant mode of transmission. The defect itself has been isolated to the *FBN1* gene on chromosome 15, which codes for the connective tissue protein fibrillin [].

Dural ectasias are well-recognized sequelae of Marfan syndrome. They are formed from a weakening or expansion of the dura, which can slowly enlarge with CSF pulsations and eventually erode through the vertebral body [6]. According to Tordis Böker, fifty-two of 58 patients with hereditary connective tissue disorders and 11 controls had dural ectasia at follow-up. Forty-five Marfan patients had dural ectasia at follow-up vs. 41 at baseline. Five Loeys-Dietz patients had dural ectasia at follow-up vs. four at baseline. Twenty-four Marfan and 2 Loeys-Dietz patients had anterior sacral meningocele at follow-up, compared with 21 and 1, respectively, at baseline [7].

Although there is no specific incidence of dural ectasia with Marfan syndrome, Mesfin A reported high frequency of dural ectasia with Marfan patients up to 89% in his study [8].

This is in accordance with our first and third cases who were sisters and diagnosed as Marfan syndrome.

Due to the fact that females undergo pelvic imaging more than males for unrelated reasons, ASM is more common in females by about three- to fourfold as

Table 1 clinical and demographic data of studied patients

Case n	Age	Sex	Complain	Motor	Sensory	Sphincter	Location	Approach	Outcome
1	20	F	Abdominal swelling	Intact	Intact	Intact	Lumbar	Anterior	Improved
2	21	F	Menstrual irregularities	Foot drop	Intact	Intact	Sacral	Anterior	Foot drop the same but she had regular menses
3	18	F	Abdominal pain	Intact	Intact	Frequency	Lumbar	Anterior	Improved

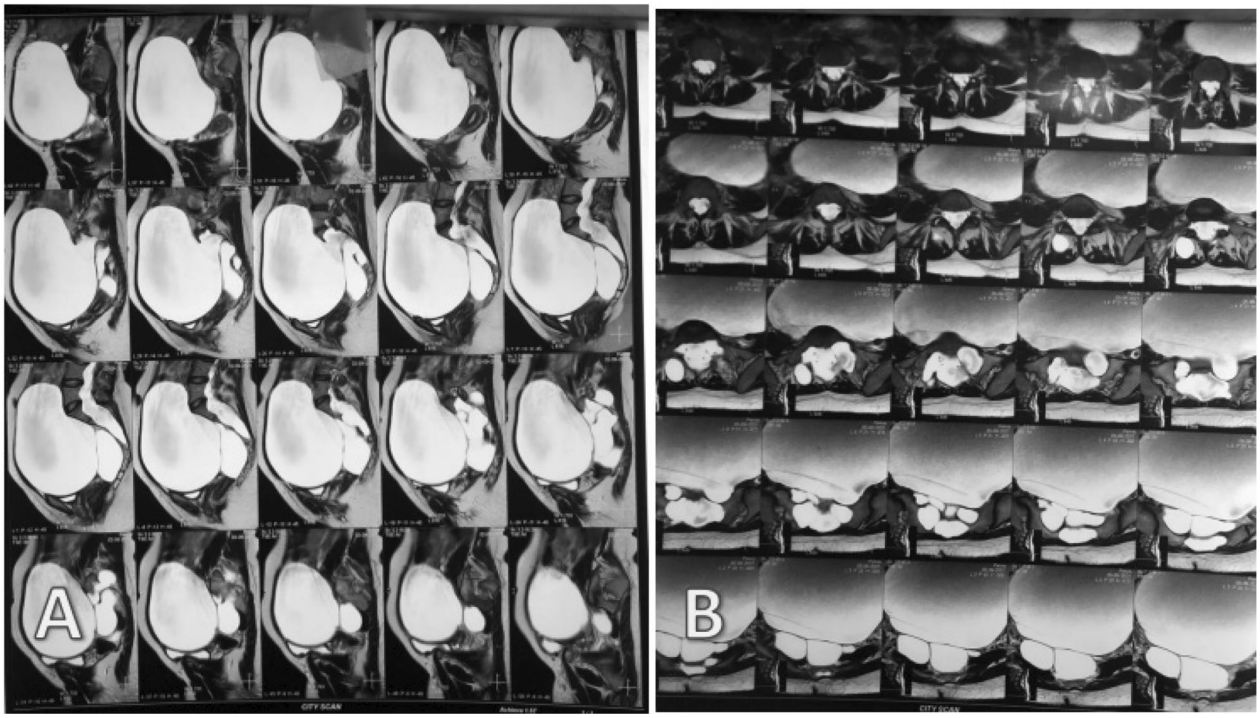

Fig. 1 Preoperative sagittal (**A**) and axial (**B**) MRI T2WI of abdomen and pelvis showing a huge anterior sacral meningocele with multiple separations

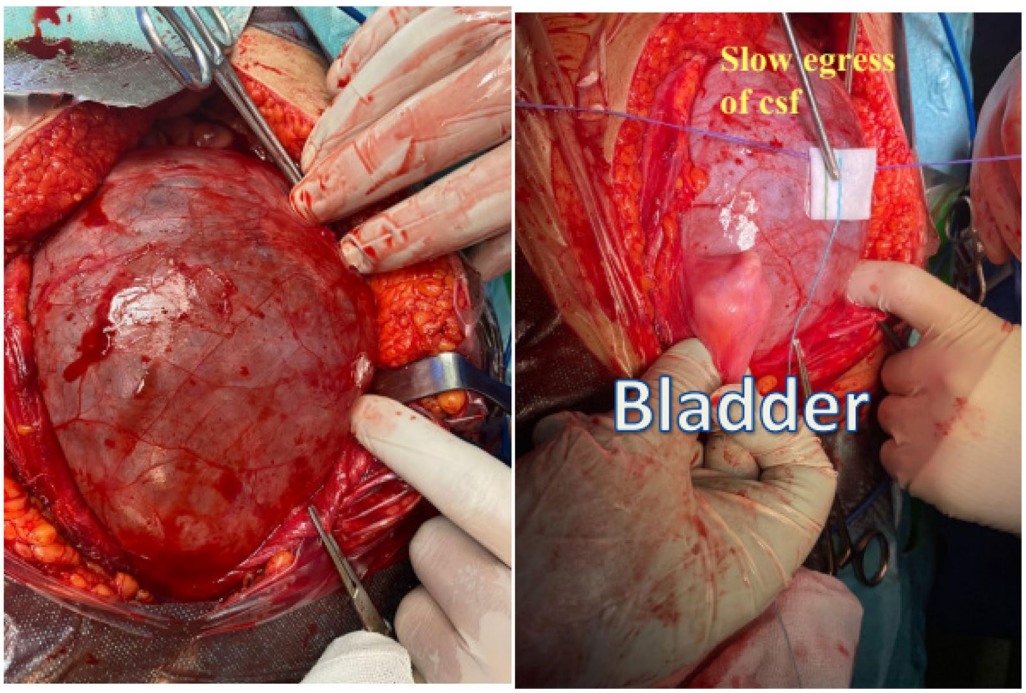

Fig. 2 Intraoperative image where the meningeal sac can be seen through an anterior abdominal incision (**A**), all abdominal contents and viscera have been shifted laterally by the lesion, a Foley's catheter within an empty urinary bladder can be seen held (**B**) next to the adherent sac

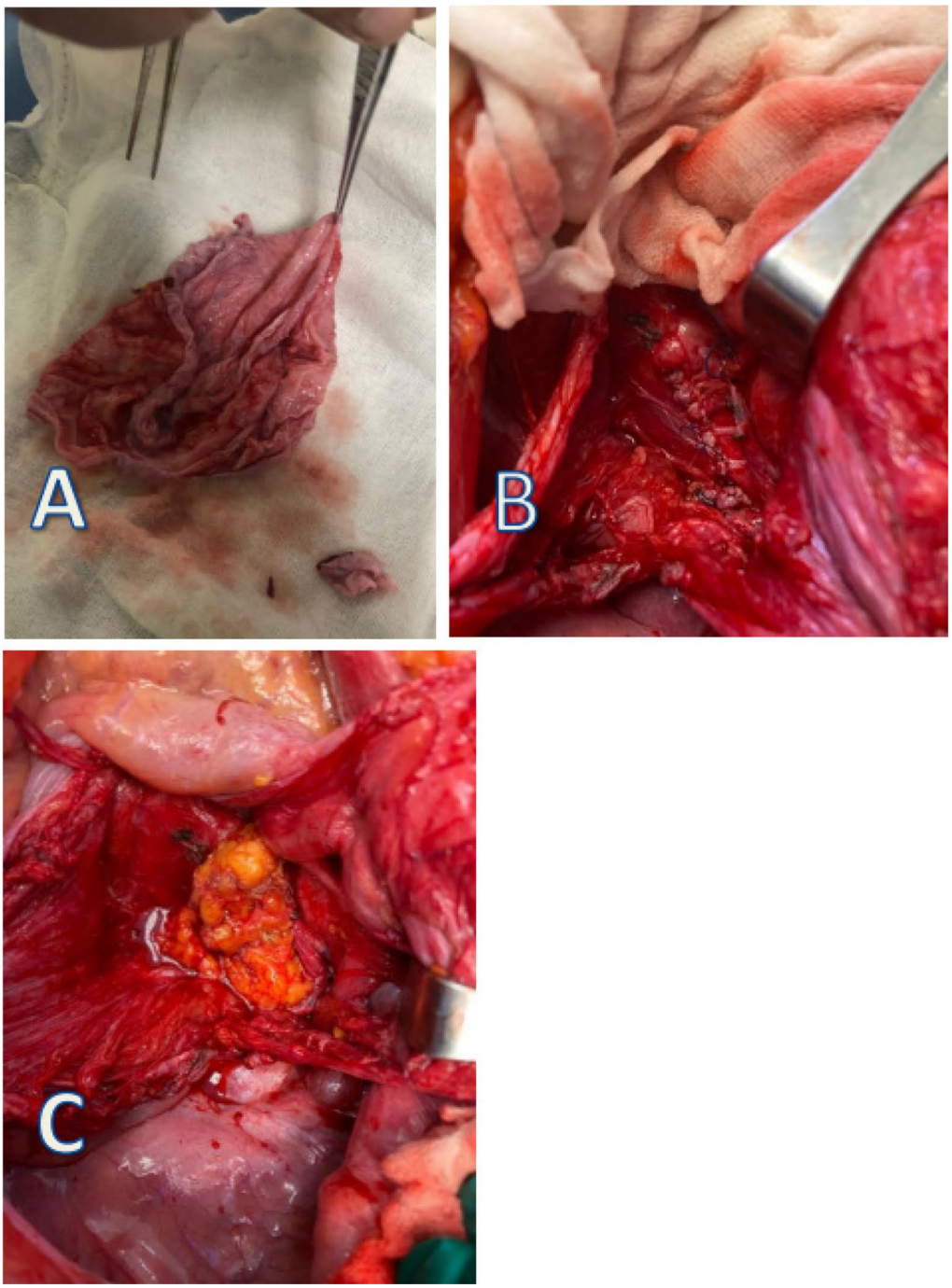

Fig. 3 Intraoperative image of the excised sac (**A**), watertight closure of the pedicle was done. (**B**) With a fat graft laid on top for additional support (**C**)

claimed by Villarejo [9]. In a study made by Cheng on 11 patients with ASM, 7 patients were females [10]. This is in accordance with our 3 cases that were young age females.

Other significant associations with Marfan syndrome are cardiac manifestations such as aortic dilatation, aortic regurgitation, aneurysms and mitral valve prolapse that requires valve replacement can occur as well. All previous mentioned cardiac manifestations are the most worrisome clinical findings [11].

So, the study recommends complete cardiac assessment in patients with Marfan syndrome preoperatively, as we did with our study cases.

Several approaches are widely described in literature regards the management of ASM including anterior transabdominal, posterior transsacral and posterior sagittal approaches.

One of the past trials was the transvaginal aspiration of the cyst but was rapidly abandoned ?? due to the high rate of complications as recurrence and infection as reported by Susan Jawad as aspiration from a pelvic cystic mass transvaginally ended in ASM abcess, because the patient was misdiagnosed as hydrosalpinx in a patient with connective tissue disease [12].

The cases of the study were females at a young age in the reproductive period, the second case presented with amenorrhea and requested a gynecological advice, and the third case consulted gynecologist for her abdominal pain, who misdiagnosed her as an ovarian cyst.

ASM should be an important differential diagnosis in females diagnosed with ovarian cyst to avoid misdiagnosis.

The posterior transsacral approach is useful in visualizing the fistula and disconnects it, leaving the sac for spontaneous resorption. But this approach may be complicated with anal stenosis due to disruption of the anal sphincter through this approach [4]. Therefore it is recommended in patients with rectoanal anomalies which can be corrected through this approach.

The previous literature suggests that posterior approach would be sufficient by interrupting the fistula and the cyst will spontaneously and gradually resolve.

However, in cases with huge cyst mass, the study recommends the anterior approach to completely excise the cyst for rapid relief of symptoms from the pressure effect, as in our cases, like urine retention or infertility.

Repair with omental flap could be done as reported by Paisan with favorable outcome on follow-up [4]. In our study of abdominal and sacral anterior meningocele, we replaced the omental flap step by augmentation of the site of repair using abdominal fat and artificial glue. This technique was done in the first two cases and showed excellent outcome regarding CSF leak as neither of the cases developed CSF leak on follow-up nor recurrence on the last clinic visit.

According to Cheng C, he operated 11 patients with anterior sacral meningocele via dorsal transsacral approach. Ten patients' symptoms were improved and one case developed epidermoid cyst which needed a second setting with laparoscopic excision by a general surgeon [10].

Our one-year follow-up showed improvement in the presenting symptoms with no dural ectasia or recurrence regarding the first 2 cases.

In huge anterior meningocele, we recommend and stress on slow egression of CSF to avoid bradycardia, hemodynamic instability or any chance of transtentorial herniation. This is also recommended by Ojeda in his literature [13]. In the present study, we did slow egression of CSF through tapping by narrow pore needle; then, a small opening was done with 11-mm scalpel and slow drainage over 20 min.

We added a preoperative step by insertion of a lumbar drain to control intrathecal CSF pressure and by gentle pressure on the abdomen we were able to increase drainage through the lumbar drain and slightly decompress the cyst before incision and removed it on second day postoperative after confirmation by abdominal ultrasound that there is no complication.

This step was described by Ashley Jr WW in his case report [14].

Although he kept the lumbar drain open for three days postoperative, we did not see any need for keeping it longer.

Conclusion

Anterior spinal dysraphism is a rare pathology with difficult diagnosis and approach. They can be asymptomatic or may manifest clinically. Symptoms are caused by their mass effect on surrounding pelvic structures resulting in chronic constipation, gynecological problems and urine problems, and other manifestations. Anterior meningocele may be operated via posterior approach or anterior trans-abdominal approach according to the size of the sac and the clinical presentation. Meticulous perioperative management with multidisciplinary team approach is mandatory to decrease morbidity.

Recommendations

- Any female with abdominal swelling with suspected Marfan syndrome or amenorrhea should be furtherly investigated to exclude ASM.
- Patient with ASM and Marfan syndrome preoperatively should be exposed to full cardiac assessment.
- Other family members should be screened for connective tissue diseases.
- The lumbar drain should be inserted preoperatively. We recommend slow egress of CSF intraoperatively.
- Direct repair with augmentation with fat and glue are important surgical remarks in dealing with these cases.

- Valsalva maneuver is recommended after completion of the repair to ensure competency of repair site.
- Postoperative abdominal ultrasound is recommended on second day postoperative to exclude neither complication nor large residual before removal of lumbar drain.
- Multidisciplinary team approach is mandatory for proper diagnosis, selection of approach and safety of patients to decrease rate of complication.

Abbreviations

ASM	Anterior spinal meningocele
CSF	Cerebrospinal fluid
MRI	Magnetic resonance imaging
CT	Computerized tomography

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Abdelhakeem A. Essa is the corresponding author. Ismail Taha helped in data collection. Doaa Wadie Maximous Dawoud helped in manuscript writing. Hussien Elkhayat assisted in manuscript revision. Mohamed Anwar helped in manuscript revision.

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Availability of data and materials
Data are available upon request.

Declarations

Ethics approval and consent to participate
All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national amendments or comparable ethical standards and approved by Institutional Board Review of Faculty of Medicine Assiut University, Egypt under number (04-2023-300281).

Consent for publication
Written informed consent was obtained from the patients for publication at this clinical series research and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declared that there was no conflict of interest.

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