Anterior sacral meningocele in pregnancy

Case report

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Anterior sacral meningocele was first described in 1837. Most reported cases were associated with complications, including meningitis and death, because of misdiagnosis or inappropriate surgical approach. The authors present a case of anterior sacral meningocele accidentally discovered during pregnancy and provide unique magnetic resonance imaging documentation. The pathogenesis, management, and surgical technique are discussed.

KEY WORDS • anterior sacral meningocele • Curra rino triad • magnetic resonance imaging • pregnancy

A NTERIOR sacral meningocele is a congenital anterior herniation of the dura mater and arachnoid either through a sacral defect or an enlarged intervertebral foramen. The pathogenesis is still unclear. Nine cases have already been diagnosed in pregnant women mostly because of obstructed labor. We present the first MR imaging illustration of ASM in pregnancy.

Case Report

This 27-year-old woman, who had been pregnant three times, given birth once, and had one abortion, presented for her routine 22 weeks’ gestation visit. Her second pregnancy ended with normal vaginal delivery, and antenatal abdominal ultrasonography findings were reported as normal. Vaginal examination and pelvic ultrasonography revealed a cystic mass. Laparotomy was conducted to locate an ovarian cyst but none was found. Postoperative MR imaging revealed an ASM.

At 38 weeks’ gestation the patient underwent an elective cesarean section birth. Repeated MR imaging (Fig. 1) at 35 weeks’ gestation had revealed no increase in the size of the meningocele. The meningocele was surgically treated via a sacral laminectomy 4 months later. Surgical exploration revealed an intrasacral meningocele communicating anteriorly and to the left with an ASM. The upper part of the opening in between showed an epidermoid cyst, which we resected. Cerebrospinal fluid was drained from the ASM and the depth of its cavity measured 7 cm.

The upper portion of the ISM and the dural sac communicated via a tiny hole surrounded by nerve fibers. Repair was performed by patching, and sutures were secured with Surgicel and fibrin sealant. Postoperative rectal examination showed total disappearance of the ASM.

The postoperative course was uneventful for 1 week. The patient then developed wound swelling, cerebrospinal fluid leak, and meningitis. At emergency reoperation we found a purulent fluid leak and dehiscence of suture lines. The wound was reclosed, and an external lumbar cerebrospinal fluid drain was placed for 2 weeks. The patient remained free of symptoms, and repeated MR imaging demonstrated progressive decrease in the size of the meningocele until it completely disappeared (Fig. 2).

Her asymptomatic 5-year-old daughter (second pregnancy) underwent radiography of the pelvis, and a scimitar-shaped sacrum was observed; MR imaging revealed an ASM. Family history indicated that our patient’s mother died of unexplained fever.

Discussion

Anterior sacral meningocele is a congenital anomaly that has probably been underdiagnosed. Between its first description by Bryant in 1837 and 1989, only 155 cases were reported. During the last decade, at least 65 cases were reported. Likely, this is due to the advent of MR imaging and the greater awareness on the part of physicians. Our case is probably the 10th ASM diagnosed during pregnancy.

The pathogenesis can be better understood in light of the associated congenital anomalies. The association of anorectal anomaly, anterior sacral defect, and anterior sacral...
mass is known as Currarino triad. Cases of ASM could represent an incomplete form of Currarino triad. The embryogenesis of this syndrome has been widely studied. Most authors consider it to be similar to the split notochord syndrome and to occur during the first 4 embryonic weeks. Some consider it to be caused by an abnormal ectoendodermal adhesion. Dias and Azizkhan have suggested that the primary anomaly concerns the development of the caudal cell mass (tail bud) leading to abnormal secondary neurulation; disrupted mesenchymal condensation around the notochord leading to sacral defect and anterior meningocele; and persistent multipotent stem cells in the tail bud leading to presacral teratoma.

Familial cases have been reported. Our case is probably an example of familial transmission. Most authors agree that there is autosomal-dominant transmission. Few others suggest an X-linked–dominant inheritance. Lynch and colleagues have suggested a gene location on 7q36. Association with neurofibromatosis should probably also orient researchers to chromosome 17.

Patients are usually asymptomatic or present with meningitis. The first demonstration of ASM on MR imaging was provided by Martin and De Latour in 1988. Today MR imaging is by far the most valuable diagnostic tool for ASM as it confirms the diagnosis, defines its communication with the dural sac, and reveals associated tumors.
Because of the risk of rupture, surgical treatment of ASM is required. Transrectal or transvaginal puncture should not be a treatment option because of the risk of meningitis and recurrence. When an ASM is discovered during pregnancy, we recommend conservative management, elective cesarean section, and then elective surgery once the pregnancy is completed; we believe there is a higher risk if the ASM were to be surgically treated during pregnancy. This meets the recommendations of Johnson and Kofinas and colleagues.

Many surgical procedures have been described for the treatment of ASM. Most neurosurgical procedures are performed via a sacral laminectomy, which provides better control of the pedicle as well as protection of the sacral nerve roots. Any associated presacral tumor could also be removed via the same approach or via an additional posterior perineal approach. Laparotomy has been used by many surgeons, but dissection of the meningocele and access to its pedicle has proven to be difficult.

Acknowledgments

The authors thank Dr. Dominique Chaumet, for referring the patient, Dr. Beatrice Carsin, for providing the MR images, and Dr. Mustafa K. Baskaya, and Mrs. Linda Hanna for their careful reading and suggestions.

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