Missed anterior sacral meningo(myelo)cele presenting with obstructive uropathy

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ABSTRACT

A 2-month-old full-term female presented with a large anterior sacral meningo(myelo)cele resulting in transient obstructive uropathy with bilateral hydronephrosis and acute kidney injury. After initial bladder decompression and surgical resection of the meningo(myelo)cele, there was spontaneous resolution of bladder function confirmed with urodynamics. Anterior spinal meningo(myelo)cele (ASM) is a rare neural tube defect that may present with urinary dysfunction secondary to compression of the bladder and sacral nerve roots or congenital defects to the bladder nervous supply. Obstructive uropathy due to ASM may spontaneously resolve after surgical resection.

Introduction

Anterior spinal meningo(myelo)cele (ASM) is a rare form of spinal dysraphism in which the meninges herniates through the anterior sacral hiatus into the retroperitoneal and presacral space. Patients may present with postural headaches, abdominal or low back pain, bowel or bladder dysfunction, meningitis, or neuropathy. Females can experience dysmenorrhea, dyspareunia, or dystocia. It typically presents in adulthood, with the majority of cases diagnosed in the third decade. Surgical resection is recommended, although the neurological sequelae may be permanent. A case of ASM with obstructive uropathy in a female infant is presented along with a discussion of implications for subsequent urinary function following surgical resection.

Case presentation

A 2-month-old full-term female weighing 6.9 kg with no past medical or surgical history presented to an outside hospital with nausea, vomiting, and straining with urination and bowel movements. Parents report that prenatal ultrasound suggested an ovarian cyst but were not told of any other abnormalities. Amniotic fluid levels were normal on prenatal ultrasound (US). No gross motor weaknesses were appreciated on physical examination. Labs were significant for elevated creatinine of 2.7mg/dL. Pyloric stenosis was ruled out by ultrasound. A renal/bladder physical examination. Labs were significant for elevated creatinine of 2.7mg/dL. Pyloric stenosis was ruled out by ultrasound. A renal/bladder

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renal bladder US demonstrated resolution of the bilateral hydronephrosis. A voiding cystourethrogram on POD 4 confirmed no evidence of vesicoureteral reflux. The patient was discharged on POD 5 with a creatinine of 0.15mg/dL.

By POD17, parents reported the patient had spontaneous voids with PVRs consistently <10mL. A urodynamics study confirmed no evidence of detrusor overactivity, detrusor-sphincter-dyssynergia, or incontinence. There was no loss of compliance with a maximum detrusor pressure of <5cmH2O. Patient was unable to void during the study. Bladder was emptied of 42mL contrast. Fluoroscopic images demonstrated a closed bladder neck and a smooth walled bladder without trabeculations. There was no evidence of vesicoureteral reflux. Since patient had clinically low PVRs, patient was taken off the CIC regimen.

Discussion

ASM is an exceedingly rare congenital neural tube defect, with fewer than 250 cases reported. More than 75% of cases reported were in females, though this has been attributed to pelvic examinations performed for symptomatic ASMs. The mesoderm that form the vertebrae also give rise to the mesonephros, which explains why many defects are associated with urogenital abnormalities. This can be attributed either to pressure on the sacral nerve roots by the meningomyelocele or congenital defects in nerve supply to the bladder. In a case review of 125 cases of ASM by Oren et al., 8.8% presented with urinary retention, 8.8% with urinary incontinence, 5.6% with urinary frequency, 7.2% with urinary tract infection, and 12% with dysuria. ASM can also cause detrusor-sphincter dyssynergia (DSD), which may result in upper tract dilation and high-pressure vesicoureteral reflux. However, the urological sequelae of NTDs can lead to significant morbidity and mortality.

Early identification of ASMs and treatment is key to prevention of permanent urological sequelae. Conservative management strategies include clean intermittent catheterization for detrusor and sphincter inactivity or oxybutynin for detrusor overactivity. This combination has been shown to dramatically improve detrusor compliance and resolve vesicoureteral reflux. Surgical repair has been generally shown to improve urological function, though the degree of improvement varies with different types of NTDs. While it is clear there is substantial variation in the urological presentation of ASM, no data currently exists on the long-term urological outcomes of ASM or the efficacy of conservative versus surgical management. Given that surgical correction of
ASM is recommended to prevent progression of neurological dysfunction, these patients should be followed postoperatively to manage any long-term urologic sequelae.

Conclusion

Anterior spinal meningomyelocele is a rare condition and can be diagnosed with appropriate history, physical examination, and adjunctive imaging. It can result in obstructive uropathy, but after surgical resection, spontaneous resolution of the bladder dysfunction can be assessed with urodynamics. Patients should be followed after surgical resection, as there may be residual long-term bladder dysfunction.

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Declaration of competing interest

None.

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