Thoracic spinal arachnoid cyst in a pediatric patient presenting with isolated bladder and bowel incontinence

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ABSTRACT

Spinal arachnoid cysts (SAC) are uncommon benign spinal cord lesions, particularly in children, that can result in a variety of neurologic symptoms, including neurogenic bladder. Here we present the case of a 7-year-old female with new onset, isolated urinary and stool incontinence who was found to have a T4-T7 SAC. Though this was initially believed to be an incidental imaging finding, after thorough work-up and persistence of her symptoms despite conservative measures she underwent neurosurgical intervention with complete resolution of incontinence. SAC represents a very rare but potentially reversible cause of neurogenic bladder that the urologist should be aware of.

1. Introduction

Spinal arachnoid cysts (SAC) are uncommon benign spinal cord lesions, that can result in a variety of neurologic conditions, including neurogenic bladder. SACs most commonly occur in middle-aged patients and arise from the thoracic spinal cord. They can cause focal spinal cord compression secondary to both the SAC itself and turbulent cerebrospinal fluid (CSF) flow around the cyst. Symptoms are typically dependent on the location of the SAC and can include pain, sensory or motor deficits and less commonly bladder or bladder dysfunction. Frequently, SACs are an asymptomatic incidental imaging finding. Here we describe a unique case of a pediatric patient with a thoracic SAC presenting with new-onset incontinence that resolved after neurosurgical intervention.

2. Case

A 7-year-old previously healthy female presented with two weeks of urinary incontinence (UI) and three days of fecal incontinence (FI). UI occurred day and nighttime with variable volumes and often without sensation prior to leakage. Labs were unremarkable and renal bladder ultrasound (RBUS) showed normal kidneys and bladder. A spinal MRI showed no lower spine abnormalities but a T4-T7 dorsal SAC with mild compression of the cord (Fig. 1). She had no sensory or motor deficits on neurological exam and given isolated incontinence and thoracic level of SAC, this was felt to be an incidental finding after discussion between both neurosurgery and urology teams.

Conservative measures including pelvic floor physical therapy (PT), treatment of constipation, and timed voiding were initiated, however, symptoms worsened with increasing incontinence requiring pull ups. She also had dribbling of urine during changes. She endorsed occasional paraesthesias in the vaginal area, hands and feet but continued to have no focal neurological deficits on examination. Video urodynamics showed a 320 cc bladder (estimated 180 cc) with low pressures, no vesicoureteral reflux (VUR), no detrusor overactivity (DO) and no leakage during the study. She had sensation to void and emptied to completion with a detrusor contraction. She did exhibit some pelvic floor activity during voiding with prolonged voiding time, suggesting bladder and pelvic floor dysynergia. PT and conservative measures were continued with excellent adherence and anti-cholinergic therapy initiated, however, symptoms persisted. She also underwent electrophysiologic monitoring under anesthesia, where motor evoked potentials and somatosensory evoked potentials demonstrated no abnormalities.

Given persistence of symptoms, a voiding cystourethrogram (VCUG) was performed which showed a 400 cc bladder without VUR or trabeculation, however, her bladder neck appeared open concerning for possible neurogenic sphincter dysfunction (Fig. 2). After multi-disciplinary discussion, she underwent thoracic right hemilaminectomy with excision of intradural SAC (Fig. 3) and recovered

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Once her catheter was removed, she had full return of bowel and bladder sensation with no further incontinence episodes and is now 8 months post-op with no return of symptoms.

3. Discussion

SAC is a rare but potentially correctable cause of neurogenic bladder in pediatric patients. The largest pediatric SAC series included 31 patients. Median age at presentation was 6.9 years, 55% were female and 68% had significant neurologic history including neural tube defects and hydrocephalus supporting previously reported associations between myelomeningocele (MMC) and SAC. Location was intradural in 18 (58%) of patients and included 4 (13%) thoracocervical, 11 (36%) thoracic and 6 (19%) thoracolumbar cysts suggesting thoracic predominance in children as is seen in adults. Sixty-eight percent of patients were symptomatic with back pain (42%), lower extremity weakness (39%) and gait instability (32%) being most common. Two patients (7%) had bladder dysfunction not further described, one with lumbosacral SAC and the other a T12-L2 SAC, both with MMC and additional neurologic symptoms at presentation. At a median 4.2 year follow up, 68% of patients had complete resolution and 19% had improvement of symptoms after surgical intervention, suggesting that SAC is a treatable cause of neurologic dysfunction, including neurogenic bladder. Idiopathic SAC in otherwise healthy pediatric patients is a rare diagnosis with only a small subset of these patients experiencing urinary symptoms and even fewer having no other neurologic findings. Evangelou et al. reviewed 21 cases of idiopathic pediatric SAC presenting at mean age 6.3 years. Eleven cases (84.6%) involved the thoracic spinal cord. Motor deficits were the most common symptom occurring in 16
(76.2%) patients with only one patient with urinary symptoms. This was an 8-year female presenting with recurrent UTIs and progressive constipation with no other neurologic symptoms found to have a T1-5 SAC. They reported 94% of patients had resolution of symptoms post-operatively including this patient.

To the best of our knowledge only three cases of isolated neurogenic bladder believed to be secondary to SAC have been previously reported. One is the case of the eight-year female described above. Boueva et al. reported a 6-month female found to have a large T3-T9 SAC after presenting with recurrent febrile UTIs. Initial RBUS revealed a large capacity bladder with poor bladder emptying and bilateral hydronephrosis. Serial imaging with RBUS and VCUG over a 2-month period revealed a progressively hostile bladder with development of VUR. Post-operatively she had improvement in bladder emptying and resolution of hydronephrosis. Satyarthee reported a 5-year-old male presenting with dribbling of urine, incomplete bladder emptying and nocturnal enuresis with no findings on neurologic examination or urologic work up found to have a S1–S3 SAC on MRI. They reported complete resolution of symptoms after surgical intervention. Though only a limited number of case reports exist, SACs have been described to occur at a variety of spinal levels with variable symptoms of neurogenic bladder without a clear correlation between lesion location and symptoms.

4. Conclusions

SACs are a very rare cause of neurogenic bladder in pediatric patients with our case believed to represent only the fourth reported case. Though case numbers are small and urodynamic and imaging findings are not available for all patients, urinary symptoms secondary to SAC do not seem to be uniform in all cases nor do they correlate with SAC location. It is important for the urologist to be aware of this unusual but potentially reversible cause of UI. SAC requires a high index of suspicion to diagnose and successfully treat, including interdisciplinary management by urologists and neurosurgeons.

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Declaration of competing interest

None.

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