Lower abdominal mass in a 16-year old adolescent: an unusual presentation of posterior urethral valves

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Abstract

Posterior urethral valves (PUV) are now commonly suspected on antenatal ultrasound, but can present with a broad spectrum of severity postnatally. Rarely, the diagnosis is missed until adolescence or adulthood when the patient usually presents with lower urinary tract symptoms. We describe an even rarer case of PUV in an adolescent who first presented with renal failure and a palpable lower abdominal mass due to urinary retention. We review the literature on presentation, natural history and outcomes of both early and late presenting PUV cases.

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Introduction

Posterior urethral valves (PUV) are now most frequently suspected by antenatal ultrasound. Postnatally, PUV can have a broad spectrum of presentation ranging from a life-threatening pulmonary hypoplasia due to oligohydramnios, to milder obstruction with few pathological signs or symptoms that may escape early detection and manifest only in later childhood, adolescence or even adulthood.[1-4] Older patients usually present with lower urinary tract symptoms, overflow incontinence, recurrent infections, or less commonly, ejaculatory dysfunction, gross hematuria and renal insufficiency.[3-7] We present the rare case of a 16-year-old adolescent who presented late with progressive lower abdominal distension, intermittent urinary incontinence and renal insufficiency.

Case report

A 16-year-old boy presented to the pediatric nephrologist with elevated creatinine and a 3-month history of increasing fatigue, weight loss, anorexia and abdominal distension. He described nocturia, day-time urinary incontinence and denied any urinary tract infections. He claimed to have been voiding "normally" throughout childhood. His past medical history only included attention deficit hyperactivity disorder.

The patient was hypertensive at 160/100 mmHg, and at the third percentile for weight. Abdominal exam revealed a large firm lower midline mass extending beyond his umbilicus. Neurologic exam and lumbrosacral spine were normal. Bloodwork showed elevated urea of 8.2 mmol/L, creatinine of 142 µmol/L, phosphate of 1.65 mmol/L and intact parathyroid hormone of 25.8 pmol/L. Urine studies were unremarkable.

An abdominal/pelvic contrast-enhanced computed tomography scan (Fig. 1) had been done by the referring physician prior to urologic consultation. This study showed bilateral hydroureteronephrosis with a grossly distended and thin-walled bladder, and dilation of the posterior urethra. No spinal abnormalities were seen. A voiding cystourethrogram (VCUG) was attempted (Fig. 2, part a); however, it could not be completed because the patient was unable to void upon bladder filling to 500 cc. A subsequent VCUG was attempted 12 hours later (Fig. 2, part b), and again the patient could not void.

Cystoscopy showed a classic image of Type 1 PUV; the leaflets were incised at the 5, 7 and 12 o'clock positions. The child was discharged with a Foley catheter for 5 days.

After 8 weeks, he continued to have high residual urine volumes, prolonged uroflow and dribbling despite normalization of his serum creatinine (55 µmol/L). Post-valve ablation ultrasound showed persistent bilateral hydroureteronephrosis. Alpha antagonists were not initiated, as the clinical picture was one of bladder atony, and not dyssynergia. Given 2 previous unsuccessful attempts at VCUG and very significant patient-related anxiety, urodynamic studies were not performed. The patient could not double or triple void consistently, and so an appendicovesicostomy was indicated to facilitate clean intermittent catheterization and improve upper urinary tract drainage. This procedure was completed without complication, and he currently catheterizes 4 times daily, with continuous overnight drainage. Ultrasounds, 2 and 4 months postoperatively, showed improvement in the degree of hydroureteronephrosis (Fig. 3).

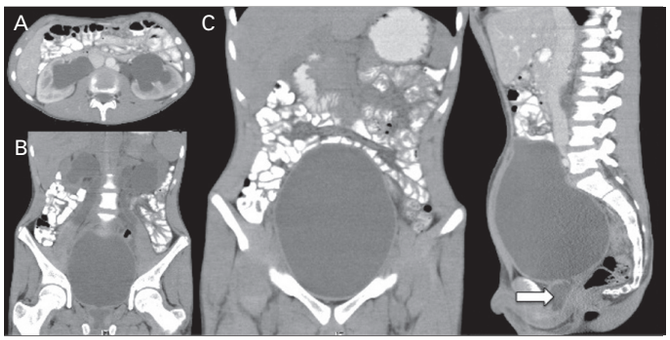

Fig. 1. Contrast enhanced computed tomography scan of the abdomen and pelvis. (A) bilateral hydronephrosis; (B) hydroureter and dilated posterior urethra ending abruptly; (C) bladder distention; (D) bladder and posterior urethral dilatation (arrow).

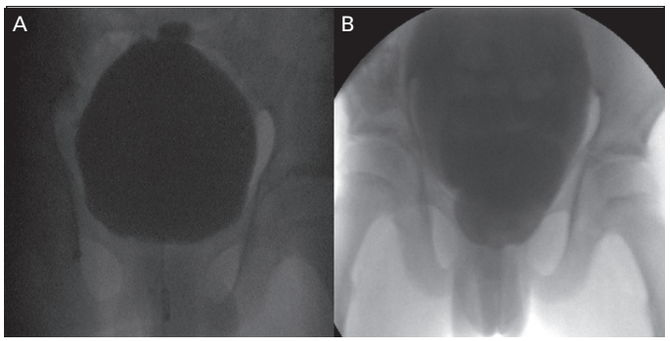

Fig. 2. A voiding cystourethrogram. (A) bladder filled to 500 cc, patient unable to void; (B) fluoroscopy 12 hours later, patient still unable to void.

Discussion

Prior to the widespread use of antenatal ultrasound, late presentation of PUV was considered a good prognostic sign, suggestive of lesser degrees of obstruction.[3] Late presentations are rare nowadays, but scattered cases have been reported in the past 2 decades.[1,3-8]

Late presenting PUV patients may display both bladder storage and emptying symptoms, incontinence, sexual dysfunction and recurrent urinary tract infections.[1,3-5] In one of the earliest and largest adult series, Mahony and Laferte studied 26 cases of PUV in men over 21 years of age. The most common presentation was frequency and urgency, followed by obstructive voiding symptoms predominantly in men older than 40 years.[4] Similar series have also found that obstructive symptoms and incontinence predominate in patients with late PUV presentation.[3,11]

Lower urinary tract dysfunction is a commom finding in boys post-valve ablation, regardless of age at presentation. Thirty-eight to 70% of neonatally diagnosed and treated PUV patients develop the "valve bladder syndrome," which represents the spectrum of bladder dysfunction that manifests many years after definitive treatment of valves.[12] Gennaro and colleagues studied the natural history of voiding patterns using urodynamic studies in 30 boys treated for PUV. Bladder dysfunction was present in 70%, and changed from unstable/hypercontractile in infancy, to hypocontractile in childhood, to a true myogenic failure in adolescence,[12] despite early valve ablation. Holmdahl suggested that these 3 urodynamic findings are variations of the same basic pattern evolving with time toward decompensation in early presenters, despite early correction of obstruction.[13] Ziyan and colleagues compared the natural history of both early and late presenters, and showed that bladder dysfunction was present in 85% of the late presenters (mean age 8.8 years); however, the same characteristics of dysfunction were also found in patients with early diagnosis (mean age 17.7 months). This similar pattern of progressive bladder dysfunction may indicate a common pathophysiology for both early and late presenters. Indeed, some suggest that the outlet obstruction during intrauterine development causes permanent changes in detrusor structure, which may be responsible for the bladder deterioration observed in adolescence.[10,11]

End-stage renal disease (ESRD) can occur in up to 43% of patients with PUV by age 30.[9] A review by Bomalaski and colleagues on 47 patients aged 5 to 35 years with delayed-presentation of PUV revealed that, at diagnosis, renal insuffiency was present in 35% and ESRD in 10%.[1] Indeed, although some suggest that late-presenting PUV carries a better long-term prognosis, renal impairment can occur at any age.

Overall, long-term outcomes after valve ablation in late presenters are variable. In their series (mean follow up 32 months), Bomalaski and colleagues found that incontinence, dysuria and weak urinary stream resolved in 18% and improved in 45%, while renal function worsened in 10%.[1] In Schober's review of 70 late-presenting PUV patients (mean age 7, range 2-14), 68% had good or improved bladder emptying immediately after valve ablation; however, 63% had ongoing daytime urinary incontinence, nocturnal enuresis or urinary frequency at mean follow-up of 25 months.[3]

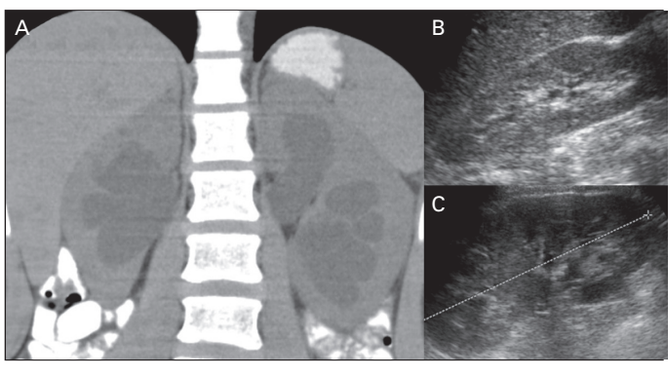

Fig. 3. A. Contrast enhanced computed tomography scan of the abdomen showing grade 3 bilateral hydronephrosis. B and C. Renal ultrasound confirming improvement in the degree of hydronephrosis.

As in our case, 80% of the patients with hydronephrosis at presentation showed no significant improvement after valve ablation. In such cases, aggressive management with clean intermittent catheterization and overnight bladder drainage should be promptly initiated to prevent further upper tract damage. Rapid improvement is usually seen, as reported in 2 recent studies.[14,15]

Conclusion

Our case highlights both the upper and lower tract effects of PUV. As outlined, bladder dysfunction in neonatally treated boys seems to evolve with age. The evolution of our patient's bladder until age 16 will remain unclear, but we suspect it went through stages of hypertrophy and overactivity, followed by complete decompensation. This unusual case underscores the spectrum of clinical findings associated with late-presenting valves. It adds to the small, but growing, body of literature on PUV presentation and emphasizes the importance of a high degree of clinical suspicion when evaluating boys of any age with palpable lower abdominal mass, bilateral hydroureteronephrosis and unusual urologic complaints.

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