CASE REPORT

Abdominal distention and renal failure in a neonate

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SUMMARY

A term male neonate presented to his paediatrician for a routine follow-up after hospital discharge. Prenatal care had been excellent and labour and delivery had been unremarkable. He had been feeding, gaining weight and was not in distress though significant abdominal distention was noted. Lab tests revealed electrolytes derangements, metabolic acidosis and renal failure. An ultrasound revealed severe unilateral hydronephrosis and echogenic kidneys. A voiding cystourethrogram revealed the definitive diagnosis which was posterior urethral valves.

BACKGROUND

This case serves as a reminder that the diagnosis of posterior urethral valves is not excluded by normal prenatal ultrasound (US) and that urinary obstruction should always be evaluated as a cause of renal failure, even in the absence of oliguria.

CASE PRESENTATION

An 8-day-old term male infant presented to his paediatrician for a routine follow-up after hospital discharge and new-onset severe abdominal distention was noted. The infant appeared well, had not been vomiting and was stooling 2–3 times/day. His mom reported changing wet diapers after every feed. He had already gained several ounces above his birth weight.

The infant's mother had a history of previous miscarriages and was followed closely during her pregnancy. She had several prenatal US, the last at approximately 28 weeks gestation, all of which were normal. The patient had two older sisters and an older brother, all of whom were healthy. There was no family history of any medical problems.

Upon examination, the infant was afebrile, and breathing comfortably with a respiratory rate of 40. Blood pressure was 82/42 and pulse was 128 bpm. Anterior fontanel was open and flat and there were no dysmorphic facies. Lungs were clear to auscultation. There was no cardiac murmur and peripheral pulses were symmetric and strong. Abdomen was tense and distended and bowel sounds were distant. Normal tanner one male genitalia were noted. Owing to concerns of an acute abdomen the patient was sent to the emergency department for further evaluation.

INVESTIGATIONS

Screening blood tests revealed the following: Na 121 mmol/l, K 6.3 mmol/l, Cl 93 mmol/l, CO₂ 12.6 mmol/l, blood urea nitrogen (BUN) 36 mg/dl, Cr 2.1 mg/dl. Blood gas showed pH 7.24, pCO₂ 25 mm Hg, HCO₃ 10.2 mEq/l and base excess 16

mEq/l. An ECG was normal. An abdominal US showed moderate abdominal ascites, a small, echogenic right kidney measuring 4.2 cm and an echogenic left kidney measuring 5.6 cm with moderately severe hydronephrosis (figure 1A,B). A voiding cystourethrogram revealed posterior urethral valves (figure 2).

DIFFERENTIAL DIAGNOSIS

Prior to knowing the results of the imaging studies a variety of diagnoses were entertained including congenital adrenal hyperplasia, inborn errors of metabolism, hypoaldosteronism, cystic kidney disease and spurious lab results. However, the lack of blood pressure instability and the presence of such a well-appearing child, without respiratory distress or any syndromic features made each of these unlikely. Once evidence of urinary obstruction was discovered, posterior urethral valves (PUV) was the only diagnosis which was consistent with the patient's age and could account for the presentation, electrolyte abnormalities and renal failure.

TREATMENT

A Foley catheter was inserted to decompress the bladder and release the urinary obstruction and a few days later the valves were surgically ablated. Sodium citrate was initiated to correct the electrolyte abnormalities and manage the type IV renal tubular acidosis.

OUTCOME AND FOLLOW-UP

With treatment, the patient's creatine improved to 0.4 mg/dl, his electrolyte abnormalities corrected and he was discharged home. Interestingly, in retrospect, the patient's mother recalled that this infant's urinary stream was not as strong as his older brother's had been.

DISCUSSION

PUV are a common cause of end-stage renal disease (ESRD) in males.¹ Many cases are diagnosed prenatally^{2–3} with the most common finding on prenatal sonogram being bilateral hydronephrosis.⁴ Other possible findings include a distended bladder and a dilated posterior urethra.⁴ Patients who are not diagnosed prenatally usually present during the neonatal period or during infancy. The initial presentation commonly includes urinary tract infection, weak urine stream or difficulty urinating,⁵ though the range of presentation is highly variable. Some patients may be asymptomatic while others may present with a toxic appearance owing to abdominal distension secondary to bladder distention or urinary extravasation.⁵ Those with urinary extravasation often have an abdominal mass on exam representing an urinoma.⁶

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Figure 1 Renal ultrasound demonstrating (A) a small echogenic right kidney measuring 4.2 cm with loss of corticomedullary differentiation and (B) an echogenic left kidney measuring 5.6 cm with severe hydronephrosis.

Electrolyte abnormalities are common in patients with PUV. Hyponatremia is usually present and is secondary to decreased Na absorption in the distal tubules.⁷ Type 4 renal tubular acidosis with hyperkalemia and metabolic acidosis^{7 8} is commonly found and is because of distal tubular resistance to aldosterone.⁹ Impaired urinary concentrating ability is also a feature secondary to downregulation of aquaporins.¹⁰

The initial management of PUV consists of correcting electrolyte abnormalities, urgently relieving the urinary obstruction and treating any infections or respiratory dysfunction. Surgical intervention has been performed prenatally in several cases, but the procedure carries considerable risk with an estimated 43% fetal mortality.¹¹ Postnatal surgical intervention is usually in the form of cystoscopic urethral valve ablation.¹² Other potential

surgical procedures include cystostomy, nephrostomy and ureteral reimplantation. After valve ablation the focus of management is on treatment of chronic kidney disease including dialysis and renal transplantation if and when the patient reaches ESRD. Another important aspect of postsurgical management is evaluation and treatment of bladder dysfunction. Urodynamic studies should be performed periodically and many patients will require intermittent bladder catheterisation and anticholinergic medications.¹³

The risk of development of ESRD in patients with PUV ranges from 13% to 64%.^{12 14–16} Several factors have been described which appear to influence the likelihood of development of ESRD in patients with PUV.^{14 17} Pohl *et al* compared a group of 19 patients with PUV who developed ESRD with a group of 23 patients with PUV who had normal estimated glomerular filtration rate (eGFR) during a median follow-up period of more than 5 years in both groups. They showed that renal volume less than third percentile, increased echogenicity and loss of corticomedullary differentiation on kidney US, more than three febrile urinary tract infections and decreased eGFR at 1 year of age are significant risk factors for development of ESRD.¹⁷

The antenatal detection of PUVs does not appear to change the risk of development of ESRD.¹⁴ One study suggested that the diagnosis of PUV after 1 year of age is associated with decreased risk of development of ESRD,¹⁸ while another study described delayed diagnosis as a poor prognostic factor.¹⁹ One possible explanation of the contradicting reports about the optimal timing of PUV diagnosis is that patients with severe disease that is not diagnosed early will have poor outcomes, whereas patients with mild disease might present later and yet have a favorable prognosis.^{14 18}

Figure 2 Voiding cystourethrogram demonstrating posterior urethral valves with an abrupt caliber change at the junction between the posterior and anterior urethra. The bladder is trabeculated. No vesicoureteral reflux was identified.

Learning points

- ▶ A normal prenatal ultrasound does not rule out the diagnosis of posterior urethral valves (PUV).
- ▶ The initial presentation of PUV varies widely and may range from essentially asymptomatic newborns to those with full-blown sepsis.
- ▶ Obstructive renal disease should always be considered in cases of renal failure, even in cases of 'normal' urine output.
- ▶ Even after valve ablation and with near-normal creatine, this child's prognosis is guarded and chronic kidney disease and even end-stage renal disease may still develop.

Competing interests None.

Patient consent Obtained.

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