Anterior Urethral Valve Presenting as Gross Hematuria in a Neonate

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Gross hematuria is uncommon in the neonatal period, with an estimated incidence of 0.21 per 1000 admissions in infants younger than one month. Although renal vein thrombosis is the most common cause, various etiologies, including congenital anomalies, must be considered. Anterior urethral valve (AUV) is a rare congenital anomaly that can cause severe obstruction and significantly impact the proximal urinary system [1].

Anatomically, AUV can cause obstruction of varying severity depending on the size and configuration of the valve [2]. The pathophysiology of AUV involves abnormal development of the urethral folds in the anterior urethra, typically at the penoscrotal junction, bulbar urethra, or penile urethra. These valves form cusp-like structures that allow antegrade flow of urine but obstruct during voiding, leading to progressive dilatation of the proximal urethra and upstream urinary tract [3].

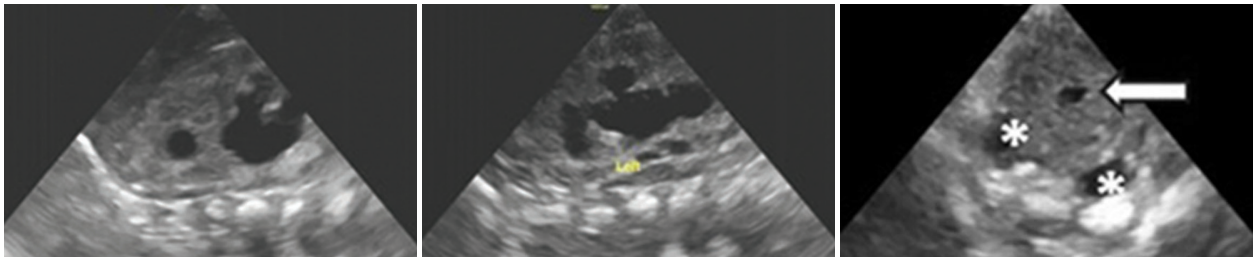
Recent data from a matched cohort study at a high-risk pediatric center found that AUV patients demonstrate significantly lower creatinine levels at initial presentation and potentially better renal outcomes compared to posterior urethral valve (PUV) patients [4]. Prenatal diagnosis of anterior urethral anomalies is feasible as early as the second trimester, with characteristic findings including anterior urethral dilation and a keyhole sign on prenatal ultrasound [5]. Unlike PUV, which are more commonly reported, AUV presenting with gross hematuria in the neonatal period is exceptionally rare, making this case particularly noteworthy for clinicians.

PATIENT DESCRIPTION

A 5-day-old male neonate presented to our pediatric emergency department with gross hematuria. The parents first noticed reddish-orange urine in the diaper a day before admission, which progressed to gross bloody urine the following morning. The baby was otherwise fine, afebrile, and breastfeeding well. Pregnancy history revealed normal standard antenatal sonography, and the infant was born at term with an

Figure 1. Point-of-care ultrasound; bilateral hydronephrosis, an empty bladder with thickened muscle and bilaterally dilated ureters

[A] Right kidney **[B]** Left kidney **[C]** *bilateral hydroureters



Arrow: thickened bladder muscle

appropriate weight for gestational age. The baby was discharged from the nursery at 72 hours of life after experiencing approximately 12% weight loss. Point-of-care ultrasound (POCUS) revealed bilateral hydronephrosis, bilateral hydroureter, and an unusually small but thickened bladder [Figure 1]. Laboratory evaluation showed acute kidney injury with a creatinine of 1.3 mg/dl (normal range for his age 0.31–0.98) and a urea level of 33 mg/dl (normal range 8–25). A complete blood count revealed a hemoglobin level of 21 g/dl without leukocytosis or thrombocytopenia. D-dimer level was elevated to 4000 ng/ml (0–500). Urinalysis revealed 3+ blood cells without leukocytes or nitrites. Further imaging, including radiological ultrasound and duplex studies, confirmed bilateral hydronephrosis with ureteral distention and no evidence of thrombosis. A computed tomography scan demonstrated moderate to severe enlargement of the collecting system and ureters bilaterally, with a non-distended bladder showing concentric diffuse wall thickening. Voiding cystourethrogram revealed a small bladder with trabeculations and a distended anterior urethra, leading to a high suspicion for an AUV diagnosis. The patient underwent a cystoscopy, which confirmed the diagnosis and urethrotomy to correct the anterior urethral valve. Postoperatively, the patient received fluid management and antibiotic therapy for post-obstructive nephropathy. The patient's condition improved with gradual weaning of fluids and catheter removal. He was discharged in good condition with normalized creatinine levels, sonographic improvement in the bilateral hydroureter, and hydronephrosis.

COMMENT

This case presents several important learning points for clinicians. First, it demonstrates that obstructive uropathy should be considered in the differential diagnosis of neonatal gross hematuria, even when antenatal sonography findings are normal. A 2024 study found that only 15.4% of AUV cases were prenatally detected compared to 72.0% of PUV cases, highlighting the diagnostic challenge of identifying AUV during routine prenatal screening [4].

The utility of POCUS in the initial evaluation of such cases is noteworthy. As demonstrated in our case, POCUS rapidly identified significant urinary tract abnormalities that raise the concern for obstructive uropathy, even before other radiology imaging studies were performed. This case also highlights the potential severity of AUV, which can cause significant upper urinary tract changes and renal dysfunction despite its anterior location. The presence of acute kidney injury, bilateral hydroureter, and hydronephrosis emphasizes the importance of early recognition and intervention. While our patient presented with gross hematuria, it is important to note that the clinical presentation of AUV is highly variable. As described in the literature, AUV may present with a spectrum of symptoms including poor urinary stream, urinary tract infections, urinary retention, abdominal distension, failure to thrive, and renal insufficiency [3]. In some cases, patients may present with a palpable penile or penoscrotal mass representing a dilated anterior urethra.

The severity of presentation often correlates with the degree of obstruction and subsequent upper urinary tract damage, ranging from asymptomatic to severe obstructive uropathy. Our findings add to the limited literature on neonatal gross hematuria as a presenting feature of the urethral valves. While most reported cases involve PUV, our case demonstrates that AUV can present in early infancy and should be included in the differential diagnosis.

CONCLUSIONS

Our case illustrates the importance of maintaining a high index of suspicion for obstructive uropathy in neonates presenting with gross hematuria, even with normal antenatal imaging. It also demonstrates the value of POCUS in the initial evaluation of such cases, potentially expediting the diagnosis and treatment.

References

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