Case - Pediatric ureteropelvic junction obstruction leading to emphysematous pyelonephritis

A rare presentation of a life-threatening condition

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INTRODUCTION

Emphysematous pyelonephritis (EPN) is a rare inflammatory condition of the kidney that develops due to chronic urinary tract infections (UTI) and urinary obstruction.[1,2] Most commonly, obstruction in EPN is due to urolithiasis or ureteropelvic junction obstruction (UPJO).[3] EPN is exceedingly rare in pediatrics, with only five reported cases in the literature.[1-5]

Pediatric EPN presents commonly with fever, nausea, lumbar/flank pain, and vital instability. Diagnosis of pediatric EPN is supported by computed tomography (CT) findings of gas in the renal parenchyma or perinephric space, often with fluid collections.[6] On CT, the differential diagnosis includes renal abscess, renal infarction, or xanthogranulomatous pyelonephritis (XGP).[6] The most commonly cultured microorganisms in pediatric EPN include *Escherichia coli* and *Proteus mirabilis*.[7]

Evidence-based management recommendations for pediatric EPN are unestablished, but principles of adult EPN treatment involve prompt intravenous (IV) antibiotics, urgent renal decompression from ureteric stent or percutaneous nephrostomy tube, and percutaneous drainage of perinephric collections.[8] For unstable patients, or patients with extensive parenchymal destruction, urgent primary nephrectomy may be required.[8]

Herein, we present a rare case of pediatric UPJO and bowel and bladder dysfunction (BBD) presenting with EPN. After initial management with ureteral stent placement, percutaneous drainage, and IV antibiotics, delayed nephrectomy was performed.

CASE REPORT

A 12-year-old female presented to the emergency department after seven days of asthenia, intermittent fevers, vomiting, and flank pain. On assessment, vital signs were stable, despite a temperature of 39.0°C, a white blood cell count of 18, and a urinalysis revealing leukocytes and nitrites. Past medical history was unremarkable.

Ultrasonography demonstrated severe left-sided hydronephrosis, and urgent ureteric stent placement was completed due to signs of early obstructive urosepsis. Intraoperative retrograde pyelogram demonstrated severe hydronephrosis with ureteropelvic junction narrowing (Figure 1). Upon ureteric stent placement, approximately 1 L of purulent and malodorous fluid was drained. A urethral catheter was placed for maximal urinary drainage.

A postoperative CT scan revealed an abnormally enlarged left kidney with severe hydronephrosis and an adjacent complex 8 cm collection with emphysematous features. Residual contrast dye from the retrograde pyelogram was visualized within the collecting system, indicating poor renal drainage in the setting of a possible underlying UPJO (Figure 2). A percutaneous drain was placed in the collection and empiric treatment with IV piperacillin/tazobactam was initiated. Urine and perinephric fluid collection results revealed *Finegoldia magna* and *Streptococcus anginosis* and antibiotic treatment was narrowed to amoxicillin.

After one week of IV antibiotics and percutaneous drainage, the patient had persistent intermittent fevers, poor appetite, and malaise. Ultrasound (US) identified residual thick-walled complex gas-containing perinephric collections. Magnetic resonance imaging (MRI) was performed to assess for additional drainage options and revealed a large bilobulated perinephric collection (10.7 x 8.2 x 4.6 cm) with mass effect on the kidney, adrenal, and spleen (Figure 3).

The parenchyma showed irregular thinning and intraparenchymal areas of necrosis and fluid locula-

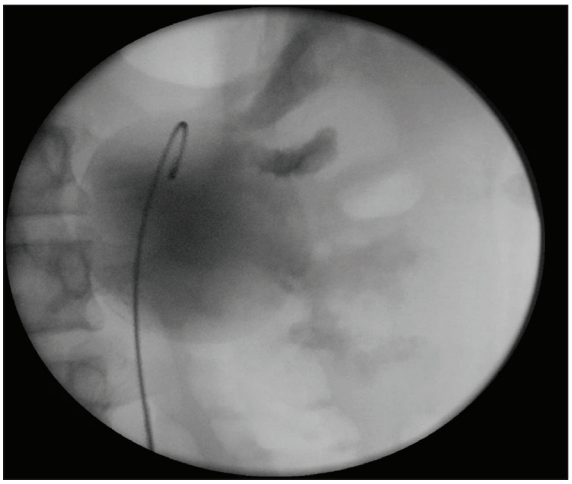

Figure 1. Intraoperative retrograde pyelogram detailing a severely dilated renal collecting system, narrowing at the ureteropelvic junction, and appropriate stent placement.

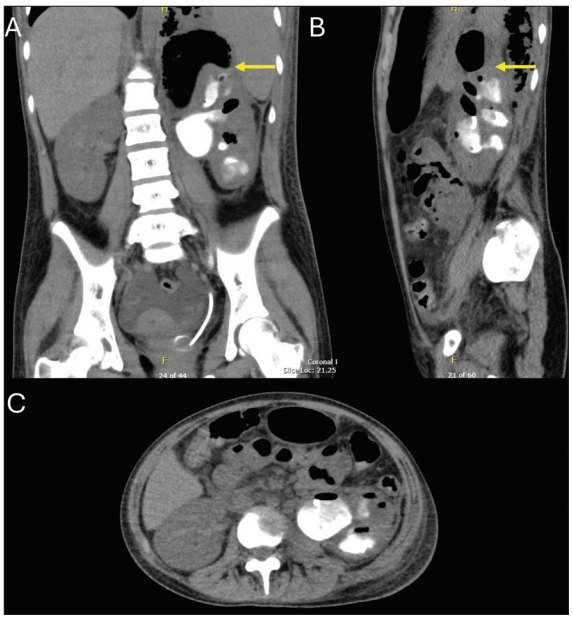

Figure 2. Computed tomography non-contrast study visualizing a dilated renal collecting system with residual contrast from prior retrograde pyelogram, signifying poor drainage. Complex gas-containing perinephric collections are noted.

tions. A dimercaptosuccinic acid (DMSA) renal scan was performed to assess for differential function for surgical planning, and demonstrated patchy cortical uptake in the upper and lower pole of the left kidney, with 10.4% split function of the renal unit (Figure 4). DMSA renal scan was performed over a mercaptoacetyltriglycine (MAG3) renogram to establish functionality; if function was preserved, MAG3 renogram was planned to determine if the kidney was obstructed and to plan for reconstructive measures and support additional drain placement. This was decided upon by

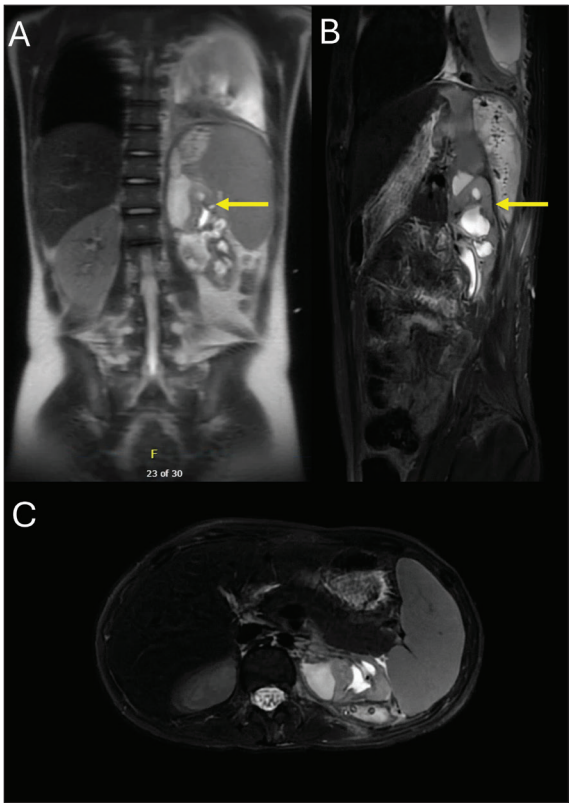

Figure 3. Magnetic resonance imaging showing persistent complex perinephric collections with mass effect on surrounding perinephric structures.

shared decision-making with the patient and patient's family.

After thorough counseling, the patient was brought to the operating room for an open nephrectomy via a subcostal approach, as there was extension of the perinephric collections toward the splenic hilum and diaphragm. One unit of packed red blood cells was administered preoperatively due to anemia. Intraoperatively, the surgical planes were obliterated with thickened and inflamed tissues. An extrinsically compressing lower-pole crossing vessel was identified as the source of the UPJO. The ureter was transected near the iliac vessels, and the stent and percutaneous drain were removed.

The surgery was performed without complications, and a drain was placed in the resection bed. Pathologic evaluation demonstrated changes consistent with atrophy and chronic infection (Figure 5). After six days in hospital, the patient was discharged home in stable condition after drain removal. A three-month postoperative US revealed resolution of the renal bed collections and a normal contralateral kidney. Symptomatically, the patient was feeling well and is now followed by

nephrology and a specialized BBD clinic for bowel management and voiding dysfunction education.

DISCUSSION

EPN is an exceedingly rare and life-threatening pediatric urologic condition. The etiology of pediatric EPN is most commonly due to chronic UTIs in the setting of obstructive calculi.[6,8] In the pediatric population, it is important to consider congenital anatomic anomalies, such as UPJO, as potential causes of EPN.[9,10]

In this case, a congenital UPJO secondary to extrinsic compression from a crossing vessel was discovered as the likely cause for urinary stasis. Concurrent intrinsic obstruction was not formally evaluated, although it was felt to be unlikely based on the retrograde pyelogram. Further investigation into the presence of intrinsic compression was not performed due to the severity of this patient's presentation. Underlying BBD likely contributed to the development of recurrent UTIs. This case highlights the importance of pediatric bowel and bladder management, as this patient's initial presentation was severe EPN.

Obliterative renal infections are rare in the pediatric literature, with XGP being more commonly reported than EPN. A review of these analogous conditions is valuable given the overlapping management.[6] Pediatric EPN/XGP presents with back pain, fever, and UTI being the most common.[1,9,11] This was consistent with our case, and exemplifies that EPN/XGP should remain on the differential diagnosis for pediatric patients with fever and back pain.[12,13]

EPN treatment should include IV antibiotics and urgent percutaneous drainage. In this case, we opted for emergent ureteric stenting due to the patient's clinical presentation, fever, and hydronephrosis seen on US. Although pediatric patients with severe EPN and/or XGP often undergo nephrectomy as definitive management, urinary and percutaneous drainage alongside antibiotic treatment was trialed to potentially avoid nephrectomy and preserve renal function.[8,12,13]

Studies have shown that antibiotic therapy can be curative for mild to moderate cases of pediatric EPN to spare renal extirpation.[8,11] Unfortunately, the patient experienced ongoing fevers and persistent complex collections despite prolonged drainage and antibiotic therapy. At this point, a thorough counseling session was completed with the patient and family.

Given the poorly functioning kidney, ongoing symptoms despite prolonged therapy, and the family's desire to proceed with definitive management to expedite recovery, we opted to proceed with a nephrectomy.

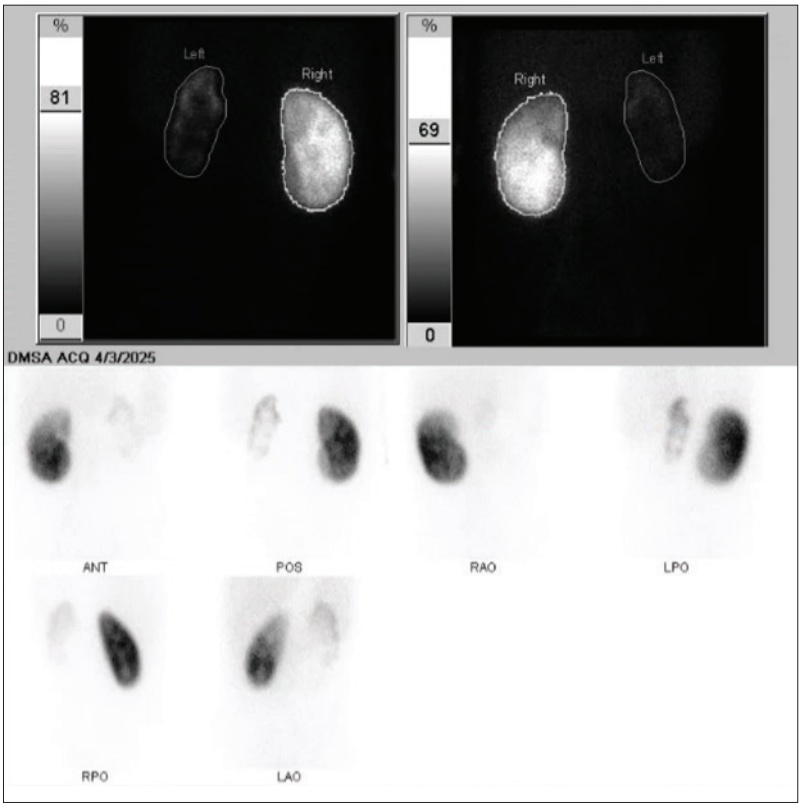

Figure 4. DMSA renal scan showing atrophied left kidney with patchy cortical uptake throughout the upper and lower pole, with 10.4% split function.

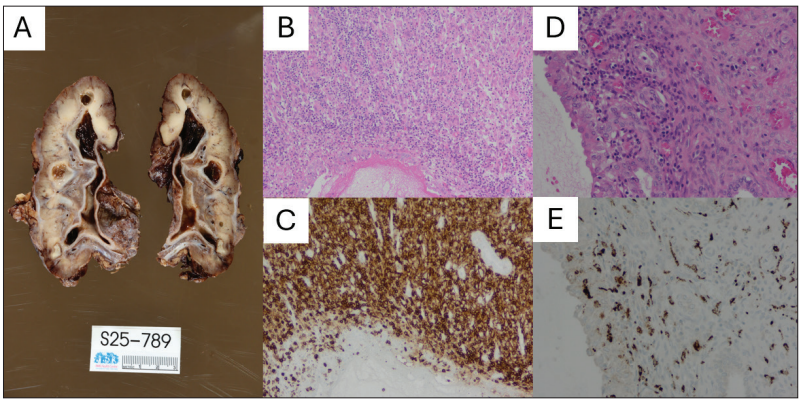

Figure 5. (A) Gross anatomy of nephrectomy specimen. (B/C) H&E and CD45 (leukocyte common antigen) stains demonstrating extensive leukocyte presence and inflammatory changes. (D/E) H&E and CD68 (monocyte/macrophage) stains demonstrating the sparse presence of histiocytes; in keeping with pathologic diagnosis of emphysematous pyelonephritis.

We believe that delayed nephrectomy with approximately two weeks of antibiotics and percutaneous drainage was beneficial for reducing postoperative complications.[8] This is in concordance with a study by Kapoor et al that demonstrated that preoperative antibiotic treatment led to reduced postoperative infection and reduced post-nephrectomy mortality, although this has not been examined in the pediatric population.[8,14]

Furthermore, the utility and timing of DMSA scans for pediatric EPN/XGP remain uncertain. In a study by Stoica et al, 90.5% of pediatric patients with XGP demonstrated <10% function, similar to what was observed in our patient.[11] It is likely that due to the late-presenting nature of pediatric EPN/XGP, most patients have already had significant renal damage, which may deem DMSA unnecessary.

The choice of DMSA vs. MAG3 renal scan was decided by a shared decision-making approach with the patient and patient's family. As the patient was having ongoing fevers, had a prolonged hospital admission, and wished to avoid additional percutaneous drains, we opted for a DMSA scan to determine functionality and to inform if nephrectomy would be an appropriate intervention compared to additional drain placement. If DMSA showed approximately equal functionality, then additional drainage would have been pursued in an effort to salvage the kidney.

Additionally, although laparoscopic approaches have been used for pediatric XGP, a subcostal approach was used in our case due to the cranial extension of the perinephric collection and the mass effect on the spleen and adrenal. Previous studies have demonstrated high incidence of bowel adherence, adhesions, and distortion of surgical planes, which increase complication rate and bowel injury.[15,16] Therefore, an open approach allowed for careful and safe dissection.

CONCLUSIONS

This case describes the rare entity of pediatric EPN treated initially conservatively with urinary and percutaneous drainage and IV antibiotics before progression to nephrectomy due to persistent symptoms. This case exemplifies the need for BBD treatment and monitoring of patients with congenital UPJO. Additionally, the role of preoperative drainage and antibiotics and the use of an open surgical approach to minimize complications were reinforced.

COMPETING INTERESTS: The authors do not report any competing personal or financial interests related to this work.

This paper has been peer-reviewed.

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