Bilateral ureteropelvic junction obstruction with ureteric stone in a female Nigerian infant: the necessity for renal and urinary tract ultrasound in acute kidney injury running head: bilateral ureteropelvic junction obstruction

Abstract

Ureteropelvic junction obstruction (UPJO) is a common cause of obstructive uropathy and can be complicated by ureteric stone and acute and chronic renal insufficiency. UPJO is commoner in male infant and is usually unilateral. We presented an eight-month-old Nigerian female infant who presented with anuria of 48 hours and in acute kidney injury (AKI). Renal and urinary tract ultrasound revealed bilateral hydronephrosis (worse on the right) with an empty urinary bladder. Urologic surgical intervention confirmed a bilateral UPJO and an impacted urinary calculus in the left UPJ. Right simple nephrectomy and left pyeloplasty with marsupialisation of the left ureteric orifice were done with improvement in renal function. She is also being followed-up for chronic kidney disease in an acquired left solitary functioning kidney. The case underscores the importance of ultrasound in the clinical management of anuric AKI in resource-constrained settings.

Keywords: bilateral ureteropelvic junction obstruction, female infant, acute kidney injury, renal calculi, ultrasound, Nigeria

Abbreviations: UPJO, ureteropelvic junction obstruction; OU, obstructive uropathy; CKD, chronic kidney disease; AKI, acute kidney injury; PUJO, pelvisureteric junction obstruction

Introduction

Ureteropelvic junction obstruction (UPJO) also known as pelvisureteric junction obstruction (PUJO), is defined as an obstruction of the flow of urine from the renal pelvis to the proximal ureter.1 UPJO is the most common cause of obstructive uropathy (OU) in children and is second only to transient, physiologic non-pathologic hydronephrosis as the most common cause of antenatal hydronephrosis.2 Its anatomic basis is from either an intrinsic or an extrinsic cause. The intrinsic obstruction result from luminal narrowing of the UPJ, with or without kinking, and is characterized by excessive connective tissue and decreased smooth muscle content of the ureteral wall.3 Extrinsic obstruction is by compression of the ureter by anomalous renal vasculature and is commoner in older children and adults.4 The incidence of UPJO is estimated to be 1 in 1500, with a male-to-female ratio of 2:1.5 The left side is commoner with a left-to-right ratio of 1:5:1.6 Bilateral cases of UPJO are also seen in 10% to 40% of neonatal hydronephrosis.7

UPJO as an OU can be complicated by sodium wasting, hyperkalaemic acidosis, nephrogenic diabetes insipidus us, urinary tract infection, failure to thrive, palpable abdominal/flank mass, intermittent abdominal pain haematuria, nephropruirous calculi, and acute renal insufficiency.7,8 However, asymptomatic cases of UPJO are possible that can be identified incidentally when the renal tract is imaged for other reasons. Concomitant lithiathosis of the urinary tract is not uncommon and whether it co-exists as a separate entity or is the result of a narrow renal outflow tract is still being debated.9 Incidences of 16% to 20% of urolithiasis have been reported on open pyeloplasty by some authors.3,9 Lampel et al also suggested that at least 14% of stones treated in such patients were associated with narrow pelvisureteric junction.10

Acute renal insufficiency in the setting of UPJO can be a form of acute on chronic kidney disease (CKD) or just an advanced CKD in an undiagnosed OU. While the two may be difficult to distinguish from the biochemistry alone, a history of an antenatal diagnosed anomaly with small kidneys or cystic kidney on ultrasound may support the diagnosis of a CKD.

We described an 8-month-old girl who presented with anuric acute kidney injury (AKI) with severely elevated azotemia but who was eventually found to have a bilateral UPJO with ureteric stone. This case illustrated the necessity of ultrasound in the evaluation of AKI as the treatment for post-renal AKI entails an urgent urological intervention. Unfortunately, ultrasound machine may not be readily available in resource-constrained settings of the world.

Informed consent

The parent of the child presented here gave an informed written permission for the publication of this case. We also obtained approval from the Health Research and Ethics Committee of the University of Abuja Teaching Hospital, Abuja, Nigeria.
Case report

An 8-month-old girl was referred from a satellite hospital to the University of Abuja Teaching Hospital (UATH), Abuja, Nigeria, with two (2) weeks history of low-grade fever, occasional vomiting and 48 hours history of failure to pass urine. No other systemic symptoms that may explain the cause of the AKI were present. For this illness, the child received anti malarial treatment and paracetamol at the referral hospital but was not exposed to gentamycin, ibuprofen or the non-steroidal anti-inflammatory medication. The child has not been treated for urinary tract infection in the past. No history to suggest polydipsia or polyuria. She has had a good urinary stream and this was the first time ever, that, the child will be presenting with anuria. The child was referred to the Nephrology Unit of the Department of Paediatrics of the UATH mainly because of the anuria.

During pregnancy, ultrasound done revealed a mild degree of hydrenephrosis but parent defaulted follow-up subsequently after delivery of the baby. There was no history to suggest oligohydramnios during pregnancy, no 77cm, and occipitofrontal circumference of 46cm. The abdomen was full at the flanks but there was no tenderness and no organomegaly. No suprapubic mass. The chest and central nervous system examination were normal. No spinal deformity or mass. She was investigated as a case of anuric AKI when bladder catheterization yielded no urine. Her evaluation included abdominopelvic ultrasound scan, serum urea electrolytes, creatinine, bicarbonate, calcium, and phosphate, serum parathyroid hormone, serum albumin and protein, liver function tests, serum glucose, full blood count including blood film for red cell morphology and malaria parasites, coagulation screen, blood culture, throat swab for culture, hepatitis B surface antigen, hepatitis C virus antibody, human immunodeficiency virus, antistreptolysin O titre, C3 and C4 complement. The serum creatinine was 320µmol/l with an estimated glomerular filtration rate of 9.6ml/minute/1.73m² from Schwartz formula, and was classified as anuric AKI “failure”, using the modified paediatric RIFLE criteria (RIFLE – ‘R’ for risk, ‘I’ for injury, ‘F’ for failure, to describe the severity of AKI; and “L” for loss of kidney function, and “E” for end-stage kidney disease, to describe the outcomes of management of AKI).

The abdominopelvic ultrasound Figure 1 showed that both kidneys were in their normal anatomical position, shapes and outlines. However, both kidneys were enlarged for age and gender, measuring 8.64 by 4.54cm for the right and 7.38 by 4.14cm for the left kidney respectively. There was marked pelvic-calyceal system dilatation which was worse on the right than the left. Note also the marked cortical thinning and increased parenchymal echogenicity with poor cortico-medullary differentiation of the right kidney.

At surgery, findings were those of bilateral UPJO, severely hydrenephrotic right kidney with very little renal parenchymal tissue. There was mild left hydrenephrosis with good renal tissue. There was an impacted stone (Figure 2) in the left UPJ and bilaterally stenosed ureteric orifices. We, however, did not have the facility to do a biochemical analysis of the stone.

She had cystoscopy with right simple nephrectomy and left dismembered pyeloplasty and marsupialisation of the left ureteric orifice. Table 1 also shows the pre- and the post-surgery serum creatinine pattern in the child. She made clear urine 3 weeks after surgery and was discharged home. She is also being followed-up in the clinic as a case of an acquired left solitary functioning kidney.
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Table 1 Serial serum electrolytes, urea and creatinine values for the child

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<th>Dates</th>
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<th>Cl (mmol/l)</th>
<th>HCO₃ (mmol/l)</th>
<th>Urea (mmol/l)</th>
<th>Creatinine (µmol/l)</th>
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Discussion

This case illustrated bilateral UPJO as an uncommon cause of anuric AKI in a female Nigerian infant. UPJO is commoner in a male gender, and attention may be deflected from it in a female infant, as we usually say “common things occur commonly”. It demonstrates the necessity of doing renal and urinary tract ultrasound scan (USS) in the evaluation of anuric AKI. While USS may be taken for granted as a routine test in the evaluation of AKI in a developed setting, unfortunately, this may not be the case in resource-constrained settings in developing countries. Herein lays the challenge as a post-renal cause of an AKI requires urgent urologic intervention as seen in the case reported. UPJO was first described as a syndrome by Dietl in 1864 and the ensuing fibrotic changes were demonstrated by Allen T.D in 1970. Subsequently, it was proven that if left untreated the narrow junction eventually leads to deterioration of renal function in the majority of cases. Concomitant lithiasis of the urinary tract is not uncommon and whether it co-exists as a separate entity or is the result of a narrow renal outflow tract is still being debated. In the case reported, UPJO was bilateral with an associated impacted urinary calculus.

During embryogenesis, the pelvic-ureteric junction forms usually around the fifth week and the initial tubular lumen of the ureteric bud become recanalized by ≥10 to 12 weeks. The pelvic-ureteric area is the last to be recanalized. Inadequate canalization is thought to be the main embryological explanation of UPJO. Extrinsic obstructions secondary to bands, kinds and aberrant vessels are also commonly encountered.

Many cases of UPJO are asymptomatic and identified accidentally when the renal tract is imaged for other reasons, in our case, because of AKI. Infants with UPJO are also at high risk of renal injury even by minor trauma. Calculi within the pelvic-ureteric junction may cause deep flank pain without radiation to the groin due to distension of the renal capsule. Calculi in the UPJ may also cause irritative voiding symptoms such as dysuria and urinary frequency. Traditionally, intravenous urogram/pyelography (IVU/P) has been performed for assessing for UPJO. Ultrasound may also show a dilated renal pelvis with a collapsed proximal ureter. In this case, there was a marked pelvic-calyceal dilatation that was worse on the right with associated marked cortical thinning of the renal parenchyma. The surgical interventions for UPJO include laparoscopic pyeloplasty, open pyeloplasty, endopyelotomy, endopyeloplasty, and robotic-assisted laparoscopic pyeloplasty.

Gladly, the renal function of the index case improved progressively after the surgical intervention. We are also monitoring closely the acquired left solitary functioning kidney at the nephrology clinic. Follow-up has included doing urinalysis for proteinuria, initial urinary tract infection prophylaxis with septrin for an upward of 5 years, serum electrolytes and creatinine, and renal ultrasound scan.

Conclusion

The clinician in a resource-constrained setting must always think about obstructive uropathy in anuric AKI, regardless of the gender of the affected child. The case presented has illustrated the possibility of UPJO to be a cause of AKI in an 8-month-old African infant.

Acknowledgements

None.

Conflict of interest

The author declares no conflict of interest.

References

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