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Pelviureteric Junction Obstruction in an Adult Presenting as a Huge Intra-Abdominal Cyst

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Abstract **Case Report**

Intra-abdominal cystic mass is common and usually presents with non-specific symptoms. When detected, it poses a diagnostic challenge due to its numerous differential diagnoses. We here-in report a 20-year-old male who presented with recurrent left sided loin pain and swelling. Abdominal ultrasound revealed a huge cystic mass in the left flank which was inseparable from the left kidney and intravenous urography confirmed the huge cystic mass to be a dilated extrarenal pelvis secondary to pelviureteric junction obstruction. Right ectopic kidney and bilateral renal malformation were also demonstrated in the patient.
Keywords: Intra-abdominal cyst, PUJ obstruction, Ectopic kidney, Renal malformation.

INTRODUCTION

Congenital renal anomalies are structural or functional defects that occur during intrauterine life and can be detected prenatally, at birth or sometimes may be detected later in early childhood. Pelviureteric junction (PUJ) obstruction is one of the commonest congenital renal anomalies which is defined as an anatomic or functional obstruction of urine flow from the renal pelvis into the ureter at the PUJ [1,2]. Generally it implies a congenital complete or partial proximal ureteric obstruction that is usually detected in utero or during early infancy. Pelviureteric junction obstruction is found in approximately 50% of foetuses with hydronephrosis, it is a disease of foetuses and early infants, it is commoner on the left side and has male preponderance with a male to female ratio of 3-4: 1 [3].

The aetiopathogenesis of congenital (primary) PUJ obstruction is not fully understood. However, various postulations have been put forward, these include; ureteral valves, ureteral hypoplasia, collagen or muscular dysgenesis, aberrant vessels and renal malrotation. The causes of secondary PUJ obstruction include; trauma, infection, proximal ureteral tumour and extrinsic compression from retroperitoneal fibrosis, and retroperitoneal tumours [1,4,5].

PUJ obstruction can coexist with other renal anomalies. The commonest associated renal anomalies include; aberrant or accessory vessels, horse shoe kidney, duplex kidney, ectopic kidney and rarely associated with extrarenal pelvis, renal malrotation and vesicoureteral reflux disease [6].

In infants, PUJ obstruction present with abdominal swelling while in older children it present with recurrent urinary tract infection (UTI), abdominal flank swelling, intermittent flank pain and or haematuria. In adults, PUJ obstruction can present with various symptoms, including flank pain, back pain, recurrent UTI and or pyelonephritis.

The roles of imaging in a patient with PUJ obstruction include; to assess renal function, determine the presence and extend of urinary obstruction, determining the cause and to detect other possible associated anomalies. Ultrasound scan (USS) and intravenous urography (IVU) are the main imaging modality of choice for the evaluation of PUJ obstruction and its associations. Other imaging of value if available is CT urography, MR Urography, diuretic renal scintigraphy and angiography.

The case reported was that of a rare case of pelviureteric junction obstruction in an adult presenting

as a huge intra-abdominal cyst with associated multiple bilateral renal anomalies.

CASE REPORT

B. A. M. is a 20-year-old man referred to the Radiology department, Federal Medical Center, Yola, Nigeria for abdominal ultrasound scan with a clinical history of progressive left flank swelling and left flank pain.

On examination there was an obvious left flank swelling which was ballotable, soft on palpation and not tender. Other systemic examinations were within normal limits. The patient's full blood count, urea, creatinine and urinalysis were within normal limits.

Ultrasound scan revealed a huge intra-abdominal cystic mass with tapering of its inferior end located in the left flank measuring 10.3 cm × 2.9 cm which represents a massively dilated left extrarenal pelvis (Fig. 1A). The dilated left extra renal pelvis was seen to contain multiple (two) floating echogenic foci casting posterior acoustic shadows, the largest measuring 1.8cm in diameter in keeping with multiple renal pelvis calculi (Fig. 1B). There was incomplete (about 90º) rotation of the left kidney with the dilated left renal pelvis displaced anteromedially. The right renal bed was empty (Fig. 2A) and the ectopic right kidney was seen in the right iliac fossa (RIF). The ectopic right kidney demonstrated absence of the central echogenic renal sinus suggestive of extrarenal renal pelvis. The ectopic right kidney was incompletely (about 90º) rotated with the renal hilum (vessels) located posterior to the renal parenchyma (Fig. 2B).

On IVU, the control film (Fig. 3A) demonstrated multiple (two) tooth-like calcific opacities projected over the left renal bed adjacent to L4 vertebral body. The post contrast images revealed both kidneys promptly and satisfactorily excreted contrast medium. The left kidney was normal in size and location but the right kidney was located in the RIF adjacent to L3-L5 vertebrae. There was incomplete rotation of the left kidney with the left calyces seen end on and the renal pelvis located anteromedially. The right kidney showed hyper-rotation with calyces seen end on with the renal pelvis located posteromedially (Fig. 3B).

There was moderate clubbing of the left calyces (grade III hydronephroses). The ipsilateral renal pelvis was markedly dilated with tapering of its lower pole given the pear shaped appearance (Fig. 4A). Significant narrowing of the PUJ was noted with significant stasis of contrast medium in the delayed images (Fig. 4B). The previously noted tooth-like dense calcific opacities were confirmed to be within the dilated left renal pelvis. The right calyces were seen end on due to degree of malrotation. The right renal pelvis was baggy in keeping with extrarenal pelvis.

Based on the above findings a diagnosis of grade III left obstructive uropathy secondary to PUJ obstruction complicated with multiple left renal pelvis calculi in a patient with bilateral multiple renal anomalies was made.

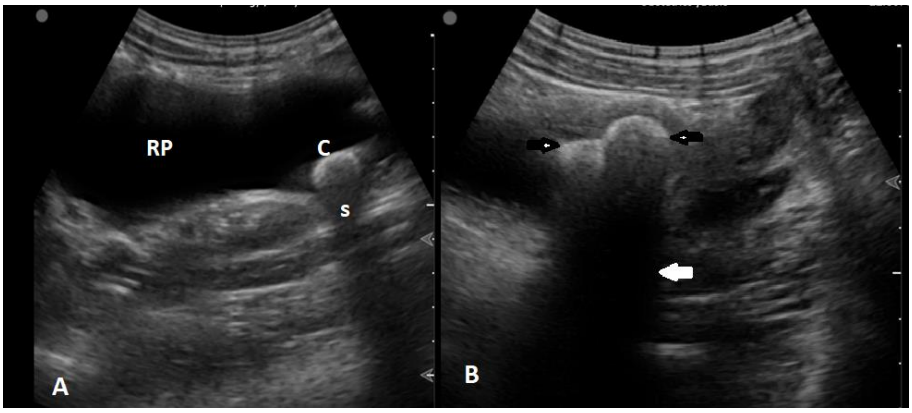

Fig-1: (A) Ultrasound images showing a massively dilated left extrarenal pelvis (RP) seen as cystic intrabdominal mass. Note the calculus (C) casting posterior acoustic shadow(s). In (B) the calculi are seen to be multiple (black arrows) casting posterior acoustic shadow (s) (white arrow)

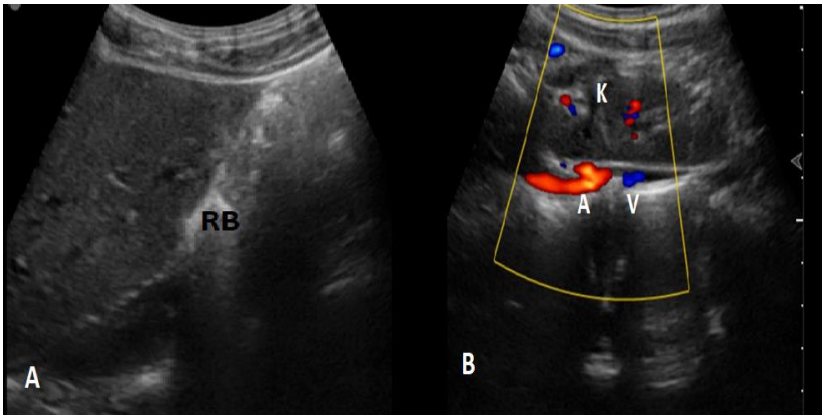

Fig-2: Ultrasound images, (A) showing empty right renal bed (RB). (B) Doppler ultrasound image showing incomplete rotation of the ectopic right kidney. Note the renal artery (A) and vein (V) entering the kidney posteriorly

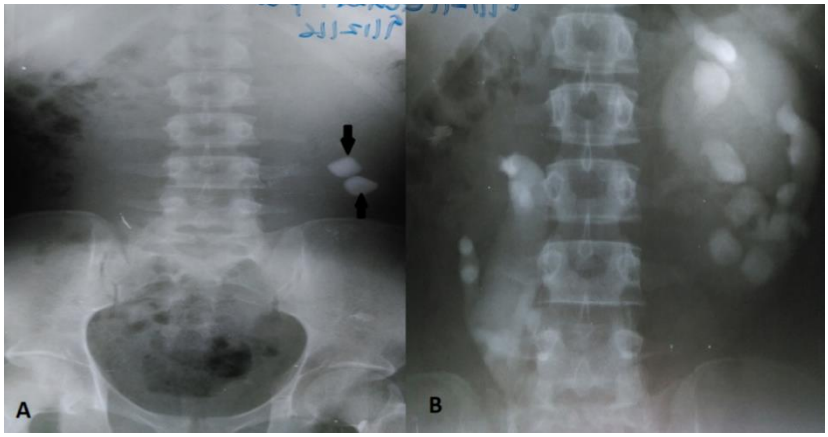

Fig-3: (A) Control film showing multiple (two) tooth-like dense calcific opacities (black arrow) projected over the left renal bed adjacent to L4 vertebral body. (B) There was moderate clubbing of the left calyces. The right calyces were seen end on due to malrotation. Note the low lying right kidney

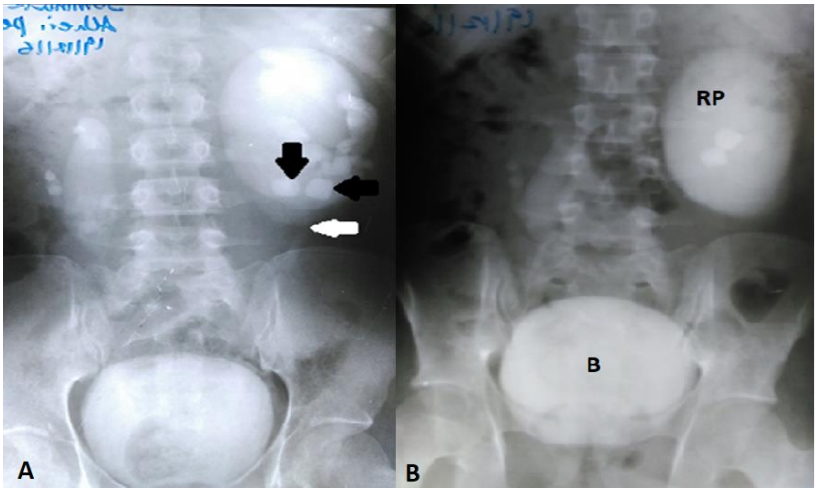

Fig-4: (A)The left renal pelvis was markedly dilated with tapering of its lower pole giving the pear-shaped appearance. (B) Significant narrowing of the PUJ was seen (white arrow). The previously noted tooth-like calculi were confirmed to be within the dilated left renal pelvis (back arrows). Significant retention of contrast medium on the 30min film of the IVU series within the dilated left renal pelvis (RP). Note the opacified urinary bladder (B)

DISCUSSION

Pelviureteric junction obstruction is one of the commonest causes of obstructive uropathy especially in infants. PUJ obstruction affects mostly the paediatric age group but can be rarely seen in adults. Some authors believe adult PUJ obstruction is under reported [7]. In children PUJ obstruction accounts for about 80% of all causes of hydronephrosis[8]. The case presented

was that of a 20-year-old patient with congenital PUJ obstruction. This might be explained by the variability in severity of the PUJ obstruction with the milder form of the disease not presenting in childhood.

More recently, smooth muscle apoptosis and defective neuronal development have been reported as other causes of congenital PUJ obstruction [9, 10]. PUJ obstruction may be associated with other renal anomalies. Das *et al.*[6] in their study reported horse shoe kidney, duplex kidney and ectopic kidney as the commonest associated renal anomalies with PUJ obstruction and the rarest was malrotation, they however did not report multiple renal anomalies in association with PUJ obstruction. The case presented had PUJ obstruction with multiple renal anomalies including bilateral renal malrotation considered a rare association. The renal malrotation might be the most probable cause of the PUJ obstruction in the index case.

Renal pelvis calculus, when it subsists simultaneously with PUJ obstruction, is presumed to be due to urine stasis with associated delayed washout of crystalline aggregates; however some authors have suggested the possibility of a fundamental metabolic abnormality as the aetiology of the stone formation [11]. This later hypothesis is further supported by the high prevalence of recurrent calculi in patients with PUJ obstruction coexisting with renal pelvis calculi. The case presented had coexisting multiple (two) renal pelvis calculi, however metabolic evaluation was not done in this patient which might have helped to determine which of the postulated causes was responsible for the calculi in this case.

After extensive review of the literature this is perhaps the second case report on PUJ obstruction with extrarenal pelvis presenting as a cystic intrabdominal mass. The earlier reported case by Johnson [10] was in a new born while in the index case is a 20 year old man and there were other associated multiple renal anomalies. The index case presented with recurrent left flank swelling, left flank pain and discomfort which are the commonest presentation of PUJ obstruction in adult.

Ultrasound scan and Intravenous urography (IVU) are the most important imaging modalities for evaluation of PUJ obstruction and its associations. However, in the evaluation of a child with a hydronephrotic kidney, diuretic renal scintigraphy has replaced IVU. The benefits of diuretic renography are that iodine-based intravenous contrast is not used, radiation exposure is minimal, and renal function can be better quantified.

Multidetector computed tomography (CT) scanning with three-dimensional reconstruction may better establish the anatomy of PUJ obstruction, extrarenal pelvis and associated vessels. Dynamic contrast-enhanced magnetic resonance urography (MRU) is the latest imaging modality used in assessing PUJ obstruction. In children, this study offers the advantages of no radiation exposure and excellent anatomical and functional details with just a single study.

The treatment options are; laparoscopic pyeloplasty or open pyeloplasty with stone extraction. The index patient had open pyeloplasty and the postoperative period was uneventful.

CONCLUSION

The case presented was that of a 20 year old man with left extrarenal pelvis and PUJ obstruction, presenting as a huge intra-abdominal cystic mass with associated right ectopic kidney and bilateral renal malrotations. The role of imaging modalities particularly USS and IVU in establishing the diagnosis was discussed. The common and uncommon anomalies associated with the condition were highlighted.

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