Case Report

New Indian Journal of Surgery
Volume 8 Number 3, July - September 2017
DOI: https://dx.doi.org/10.21088/nijs.0976.4747.8317.30

Pelviureteric Junction Obstruction in Crossed Fused Ectopia Presenting as Peritonitis: Case Report and Review of Literature

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Abstract

Pelvi-ureteric junction obstruction (PUJO) is commonly seen in infants and children. Rarely it may go undetected and presents in adults with either minimal symptoms or is detected incidentally. We report the case of a previously healthy lady aged 26 years who presented with acute abdomen, respiratory distress and shock mimicking peritonitis. During resuscitation, a bedside percutaneous abdominal drain was placed in view her poor general condition. The patient improved dramatically following drainage of intra-abdominal collection and an emergency abdominal exploration was deferred for further diagnostic evaluation. Imaging findings suggested a massively dilated crossed fused ectopic left kidney with PUJO and drain in renal pelvis. A dismembered pyeloplasty was performed electively and the patient made an uneventful recovery.

We present the clinical profile and imaging characteristics of this unknown presentation of PUJO with review of unusual clinical presentation of PUJO in adult patients from published literature. We also emphasize that this presentation of PUJO be entertained when dealing with a patient of acute abdomen and shock.

Keywords: Pelviureteric Junction Obstruction; Peritonitis.

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Received on 23.05.2017, **Accepted on** 13.06.2017

Introduction

The overall incidence of PUJO is 1:1500 with an equal male to female ratio. In nearly two-thirds of patients left kidney is affected while involvement may be bilateral in 10–46% of cases [1].

It is commonly seen in children, but may remain asymptomatic and hence undiagnosed till adulthood. Hydronephrosis picked up on antenatalultrasound (USG) scan can suggest the diagnosis of congenital PUJO antenatally. Failure to thrive prompts a suspicion for this etiology in the neonates. Children with PUJO may present with episodes of pain in flank, nausea, vomiting, gross hematuria (following minor abdominal trauma) and urinary tract infection (UTI).

Adults with PUJO may present with varied symptomatology depending on whether the resultant hydronephrosis is acute or chronic.

In PUJO the hydronephrosis is usually slow and progressive resulting in heaviness or discomfort in flank, minimal or no pain.

At times PUJO is picked up in a patient with urinary tract obstruction or acute pain secondary to spontaneous calyceal or forniceal rupture.

Acute pain may also be precipitated by increased fluid intake or ingestion of a food with diuretic properties prompting Dietel's crisis [2,3].

However, PUJO presenting as acute abdomen and shock is unheard of. We share our experience of a young lady who presented with acute abdomen and shock secondary to PUJO, the clinical course and final diagnosis surprised us. The etiopathogenesis is discussed with review of relevant literature.

Case Report

A lady aged 26 years presented in surgical emergency with chief complaints of sudden onset pain in abdomen, respiratory discomfort and altered consciousness of 1 day duration. She was previously asymptomatic and had married a month back. The pain was acute in onset, started from lower abdomen, was persistent and gradually progressed to whole abdomen within 2 hours. There were no other complaints. There was no history of fever, bowel disturbances, drug intake, trauma, any other chronic illness or previous episodes of abdominal pain. On examination, her heart rate was 130 beats per minute, blood pressure was 74/40 mm of Hg, respiration was labored with a rate of 28/ min. Her GCS was 11 (E3V4M4) with cold clammy extremities. The abdomen was tense and distended with diffuse tenderness, guarding, dull note on percussion and absent bowel sounds. A clinical diagnosis of enteric perforation or acute pancreatitis was made and the patient was resuscitated. Supplemental oxygen was started and 2 liters of warm lactated ringer was administered. Urinary catheter drained 10 ml of dark colored urine. Urine pregnancy test was negative. Arterial blood gas (ABG) analysis was suggestive of metabolic acidosis. Serum lipase, amylase and basic emergency investigations were sent. A diagnostic ascitictaprevealed turbid fluid. The patient responded partially to these interventions. In view of poor general condition, a bedside per-cutaneous abdominal drain was placed to relieve the respiratory distress. It drained 2500 ml of clear fluid with some flecks that subsided the abdominal distension completely. Further abdominal examination revealed mild diffuse tenderness; no lumpwas palpable. Patient was started on broad spectrum intravenous antibiotics and was monitored closely. Patient's general condition improved markedly following percutaneous drainage. Her central venous pressure of 6 cm saline improved to 10 cm saline after another fluid bolus of 1 liter. Routine investigations suggested hemoglobin of 9 gm%; total and differential counts, serum amylase and lipase were within normal limits but serum creatinine was 10 mg%. A repeat ABG showed trends towards normalization. There was continuous drainage of small amount of serous fluid from the abdominal drain, however the urinary catheter did not drain anything. A dramatic improvement in patient's clinical condition prompted us to defer laparotomy. Hemodialysis was done in view of deranged renal function. There was no output from the urinary catheter even after dialysis. An USG abdomen showed a little intra-abdominal collection, both kidneys were not visualized but other abdominal viscera were normal. Meanwhile, the drain output was continuous and clear. Drain fluid urea and creatinine were performed that confirmed its urinary nature. The clinical condition of the patient improved steadily and she was allowed orally. Urine routine examination excluded proteinuria. The serum creatinine level dropped from 10 mg% to 2 mg% in 48 hours. CT urography was performed that showed a large hydronephrotic left kidney with another small moiety lying below it. There was no evidence of parenchymal rupture. The drain was seen in between the two moieties. Ureter and bladder were not visualized. There was no evidence of renal calculus [Figure 1].

Cystourethroscopy was unremarkable except for a single left ureteric orifice. A retrograde pyelography (RGP) showed single left ureter with stricture in its upper end through which dye was spilling into a large confined space (presumably dilated renal pelvis) [Figure 2]. A drain contrast study showed it to drain in a closed dilated space.

A CT-RGP correlation suggested a diagnosis of fused crossed left kidney with PUJO. By this time, the patient had made complete recovery with no signs of uremia or sepsis. A plan of exploratory laparotomy was made. Intra-operatively, a visceral mass was found densely adherent to the anterior abdominal wall in the left lower abdomen. Careful dissection revealed a massively dilated left kidney with drain coursing through the sigmoid mesocolon into the huge renal pelvis. Part of the kidney, sigmoid colon and mesocolon adhered together were forming the mass. Ureter was identified with the help of uretericca the terinserted preoperatively. There was no evidence of abdominal tuberculosis. A dismembered pyeloplasty was performed with

Table 1: Unusual presentation of PUJO in adults

Author	Year	Age (yrs)/Sex	Presentation
Goldberg SD[11]	1998	31/M	Testicular pain
Uzzo RG[12]	1994	26/F	Duodenal obstruction
Tebyani N[13]	1999	65/F	Early satiety and weight loss

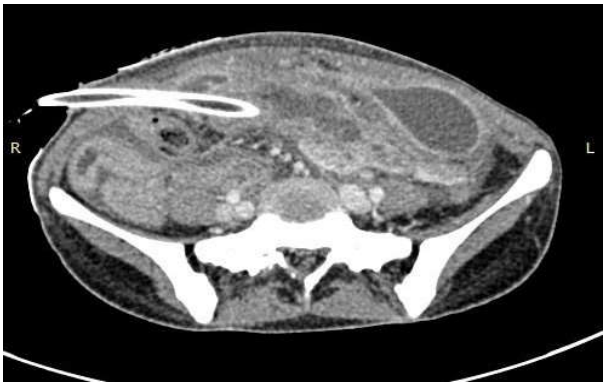

Fig. 1: Large hydronephrotic left kidney with drain in situ

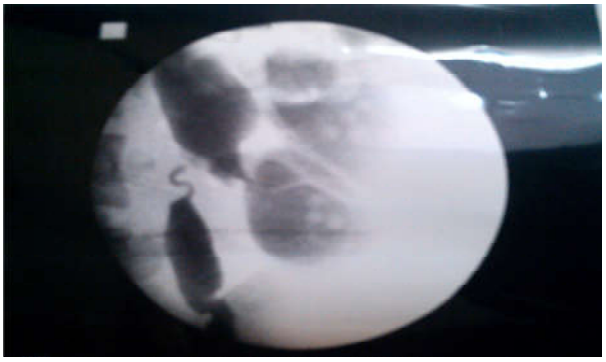

Fig. 2: RGP showing PUJO through which dye is spilling into dilated renal pelvis

uretericstentand nephrostomy. The patient made an uneventful recovery and is doing well at one year of follow up.

Discussion

PUJO is a well-known clinical condition that is caused by a variety of congenital or acquired process. Whatever the etiology, the end result is impairment of urinary drainage across the obstructed segment leading to slow, progressive deterioration of renal function over time. In an adult, usually the condition is brought to notice for evaluation of dull persistent pain or discomfort, renal insufficiency or incidentally while being evaluated for some unrelated symptom.

Rarely PUJO may lead to sudden obstruction resulting in rapid deterioration of renal function [2,4-6]. The cause of such sudden obstruction is a matter of conjecture and various hypothesis are suggested to explain this phenomena. An altered PUJ resulting in a box shaped pelvis with repeated episodes of diuresis has been cited as one of the cause for accelerated hydronephrosis [2]. This could have been the cause in the present case but there was no history suggestive of diuresis. It is possible that the excess urine produced might not have found access through the narrowed PUJ resulting in rapid hydronephrosis. This 'internal diuresis' resulting in rapid massive hydronephrosis may explain sudden severe pain and features of peritonism in our patient as is described in acute urinary obstruction secondary to calculus [7]. Peritonism coupled with the circulatory shock in our patient prompted us to consider diagnosis of bowel perforation, which is a common surgical entity. The cause of circulatory shock in our patient still eludes us. It is possible that sudden enlargement of kidney might have caused abdominal compartment syndrome leading to compression of inferior vena cava thereby reducing preload leading to shock. This mechanism has been documented in patients with bowel obstruction [8] and other space occupying disorders of abdomen [9]. This is further fostered by the dramatic improvement in the clinical condition of the patient following decompression by drain placement.

As is suggested by some authors [3,10], contralateral nephrectomy increases the excretory load of the remaining kidney that may also give rise to accelerated hydronehrosis, our patient had a fused kidney thus the excretory load on the single moiety with a small deformed outlet might have also contributed to sudden hydronephrosis. Urushibara described a case of rapid deterioration of hydronephrosis in an adult patient secondary to an aberrant vessel [5], however this was not the cause in our patient.

Although many atypical presentations of PUJO have been described by various authors (Table 1), the presentation in the current case was markedly different. Most startling was the associated shock state that lead to a diagnostic misadventure. To the best of our knowledge no similar case of PUJO has been reported with this unique symptomatology. We propose that this presentation of PUJO should be considered as a possibility in a patient with acute abdomen and shock.

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