

Crossed fused renal ectopia (pancake type) with single ureter: A case report

Kaleab Habtemichael^{a,*}, Seid Mohammed^a, Biniyam Beyene^b, Fitsum Gebreegziabher^a, Mohammed Abdulaziz^a

^a Department of Surgery, Urology Unit, St. Paul's Hospital Millennium Medical College, Addis Ababa, Ethiopia

^b Department of Radiology, St. Paul's Hospital Millennium Medical College, Addis Ababa, Ethiopia.

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ABSTRACT

Crossed fused renal ectopia is a rare congenital anomaly of renal embryogenesis. The majority of such anomalous kidneys are supplied by a single renal artery and drained by two separate ureters. However, drainage by a single common ureter is an unusual variant. Here, we present a 17-year old male with a pelvic pancake kidney drained by a single ureter with associated acute obstructive uropathy diagnosed with ultrasonography and computed tomography urography (CTU). We describe the anatomical peculiarity and diagnostic evaluation of the case.

Introduction

Fusion anomalies of kidneys are uncommon. Crossed-fused renal ectopia is the second most common fusion anomaly after Horseshoe kidney with an incidence of 1:300–7500 and a male predominance. It can present in different fusion patterns and its extreme form is pancake kidney (PK), which is described as a mass formed by complete medial fusion of renal parenchyma without an intervening septum. Each kidney often has separate collecting system and ureter.¹ Here, we present a 17-year old male with PK drained by a single ureter.

Case presentation

A 17-year old male presented to the emergency department complaining absence of urine for 2 days and a dull aching lower abdominal pain radiating to the lower back. He reported a progressive decrease in urine amount for 2 weeks before the complete absence. He had two episodes of vomiting, low-grade fever, and progressively increasing lower abdominal swelling. According to his parents, the patient had neither urinary complaints nor other illnesses during childhood.

Further examination revealed a heart rate of 108 and an axillary temperature of 38 °C. There was 10 × 8 cm firm, globular and tender swelling extending from the pubic area to just below the umbilicus. Its lower margin extends deep into the pelvis. Percussion note was dull over the mass and tympanic over the rest of the abdomen. Genital examination showed normal genital development and pubic hair distribution.

Laboratory workup revealed a white cell count of 22,000 and serum creatinine of 1.77 mg/dL. Urine dipstick showed +2 leukocyte esterase and positive nitrite whereas microscopy showed many bacteria. Ultrasonography (US) of the abdomen reported the absence of both kidneys from their respective anatomic locations. They were fused at the pelvic inlet. There was a significantly dilated renal pelvis and moderate hydronephrosis with echo debris but there was no report of hydroureter. On cystoscopic evaluation, a solitary ureteral orifice on the left side of the trigone with no visible jet of urine seen and bladder dome was indented inwards. A double J-stent was inserted through the solitary ureteric orifice without difficulty to relieve the obstruction and pain. A cystourethrography (CUG) was done and showed a severely dilated ectopic renal pelvis just above the urinary bladder and was outlined by a refluxed contrast material on the left side (Fig. 1).

A Computed Tomography Urography (CTU) performed one week later revealed a medial parenchymal fusion of pelvic ectopic kidneys that are located to the left of the midline. They had separately visible collecting systems and good power of contrast uptake and excretion (Fig. 2A and B). A significantly dilated extra-renal pelvis is seen draining both collecting systems and emptying into a single ureter. The stent is visible coiling in the pelvis proximally and left postero-lateral aspect of the urinary bladder distally (Fig. 2C and D).

The arterial supply is a single artery from the abdominal aorta at its bifurcation. The venous drainage of the right and upper moiety is to the confluence of the common iliac veins. The left and lower moiety drains to the inferior vena cava posterior to aorta (Fig. 3). The patient's clinical

* Corresponding author. Urology Unit, Department of Surgery St. Paul's Hospital Millennium Medical College (SPHMMC), PO Box 1271, Addis Ababa, Ethiopia.
E-mail addresses: kaleab528@gmail.com, kaleab528@gmail.com (K. Habtemichael), seidm96@yahoo.com (S. Mohammed), biniyam.beyene@sphmmc.edu.et (B. Beyene), fitsum1206@gmail.com (F. Gebreegziabher), ibnuabdulaziz2@gmail.com (M. Abdulaziz).

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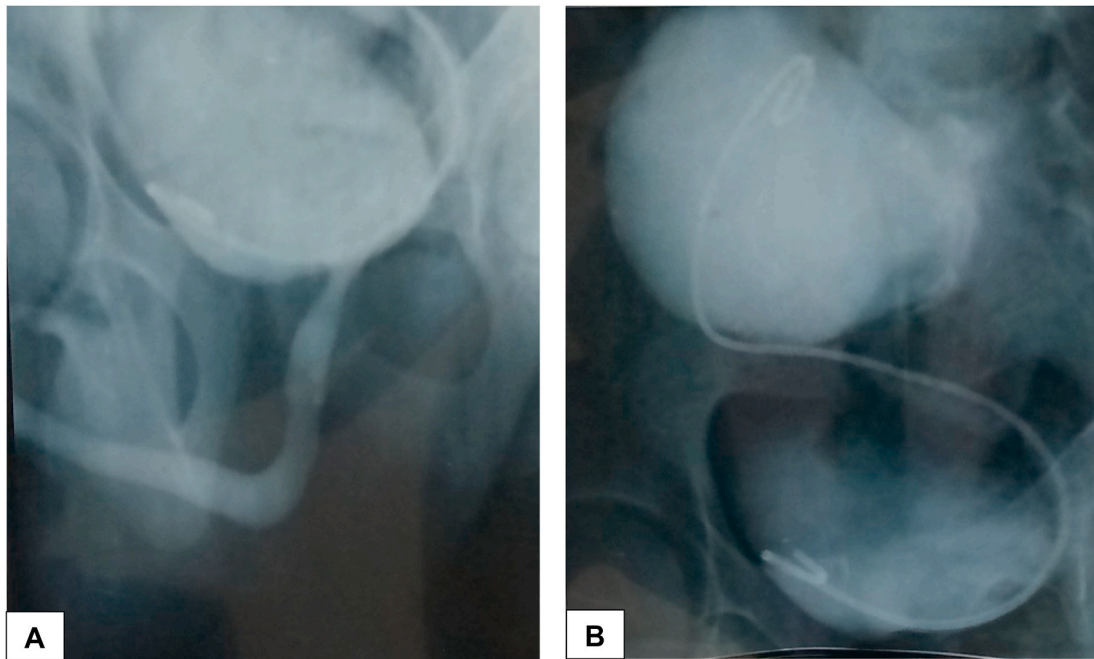


Fig. 1. Cystourethrography (CUG) which was done few days after stent placement shows a patent urethra, normal contrast filling of the urinary bladder and visible distal coil of the double –J stent (A). Reflux of contrast material into a significantly dilated renal pelvis, located just above the urinary bladder, is also seen (B).

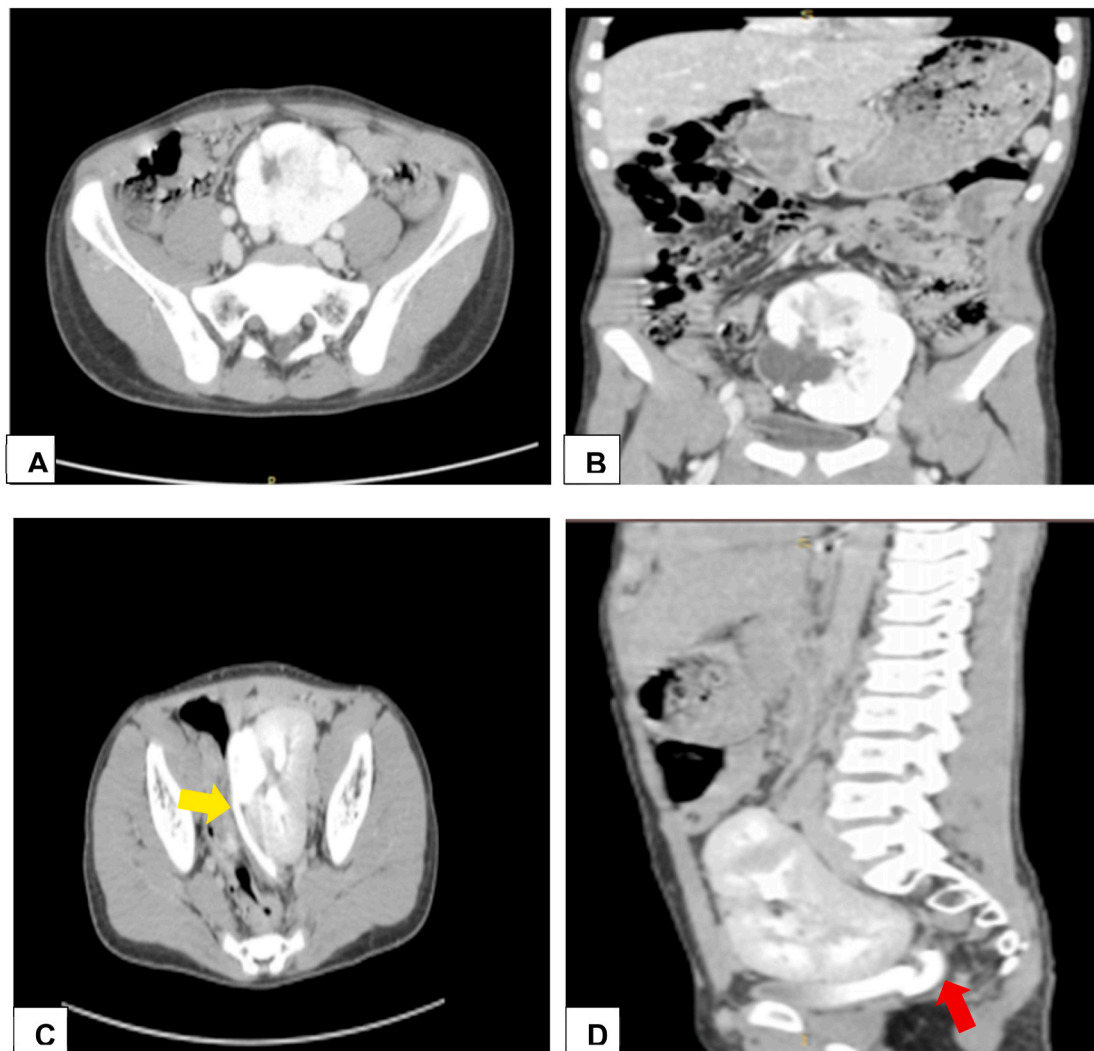


Fig. 2. Axial, Coronal and Sagittal images of CTU depicting absence of kidneys from their normal anatomic location and a pelvic pancake kidney with a good contrast uptake and a dilated extra-renal pelvis (A and B). A single ureter with a double-J stent is seen draining the renal pelvis (yellow arrow) and terminating at the urinary bladder (red arrow) (C and D). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

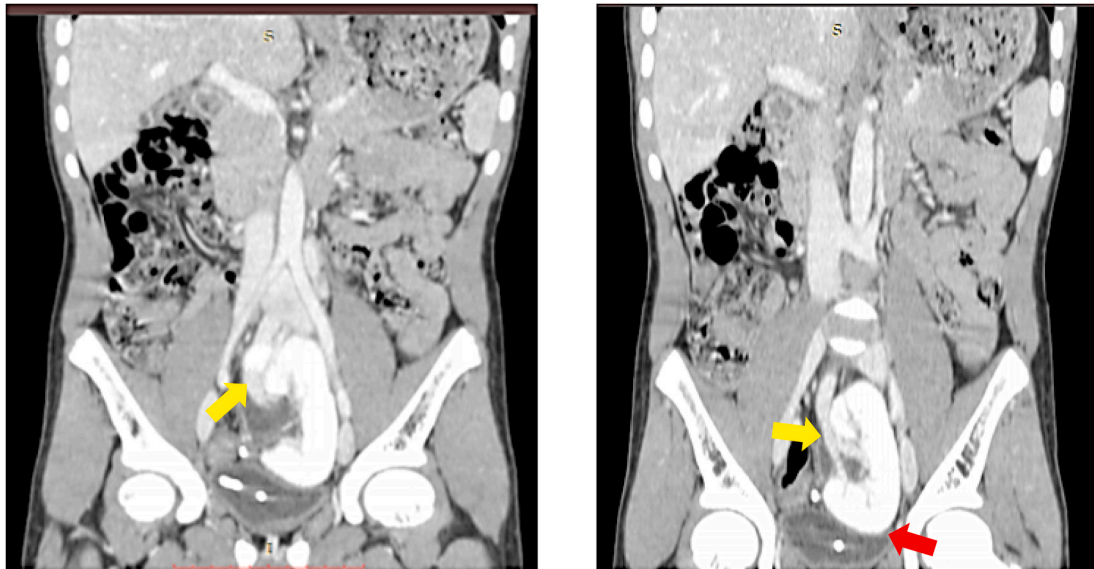


Fig. 3. Contrast-enhanced images showing a single arterial supply to the PK arising from the bifurcation of the abdominal aorta (yellow arrows). The lower border of the kidney is seen indenting the dome of the urinary bladder (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

condition subsequently improved with normalized serum creatinine (0.84 mg/dL). An exploratory laparotomy with possible pyeloplasty was planned but the parents of the patient could not give consent. Hence, patient is discharged with appointment and stent removal is planned on third month.

Discussion

Fusion anomalies of the kidneys are not rare. Several types of fusion are reported in the literature. Among these, horseshoe type is the most common (1:400). Crossed fused renal ectopia is the second most common one (1:1000–1:7500). It is formed by a medial fusion of the kidneys after one of them crosses the midline. There are few variants of crossed fused renal ectopia with PK being the rarest. Glenn et al. (1958) first used the term to describe renal fusion into one mass that is drained by two separate ureters.¹ Our case is rare because of the presence of a single ureter draining the fused kidneys and there are only a few similar reports in the literature.

Most PKs are diagnosed incidentally with only few patients developing symptoms like lower abdominal pain and hematuria.² Our patient had urosepsis and acute kidney injury. The hydronephrosis was attributed to a possible primary ureteropelvic junction obstruction (UPJO) and/or ureteral angulation due to its distorted path. Other suspected causes of the obstruction include aberrant vessel, retained mucosal folds and Fibroepithelial polyp at UPJ.

Ultrasonography and a contrast CT scan are the preferred modalities of diagnosis of PK and other associated anomalies, the common ones being imperforate anus, skeletal defects, and cardiovascular defects. Reports of unicornuate uterus, renal vascular anomalies, and UPJO also exist.³ In this regard, we did not find such anomaly in our patient.

The embryogenesis of PK is not yet well described but has been postulated in the literature that renal fusion occurs when the kidneys are pushed towards each other because of a compressive force applied as they ascend between umbilical arteries during their development. However, this theory does not fully describe the reason behind the drainage of PK by a solitary ureter and failure of a second ureter to develop.⁴

PK may not necessarily have poor prognosis. However, serious functional and infectious morbidity may arise due to associated malformations such as UPJO. In such instances, urgent relief of obstruction

with subsequent plan to correct the concomitant anomaly is required. Most reports on PK suggest a conservative management approach with regular follow-up for asymptomatic patients.^{4,5}

Conclusion

This case indicates that although PK is often diagnosed incidentally, it can also present with serious complications like renal failure and sepsis, which require urgent decompression. As there is no specific symptom of the disease, having a high index of suspicion is an essential tool in the diagnosis particularly in patients with pelvic mass and recurrent urinary tract infection. Regular follow-up visits are recommended due to the risk of calculi, infection, and obstructive nephropathy.

Availability of data and materials

The datasets with more radiologic images are available from the corresponding author upon reasonable request.

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Declaration of competing interest

The authors declare that they have no competing interests.

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