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A rare case of supernumerary kidney with crossed fused ectopia kidney

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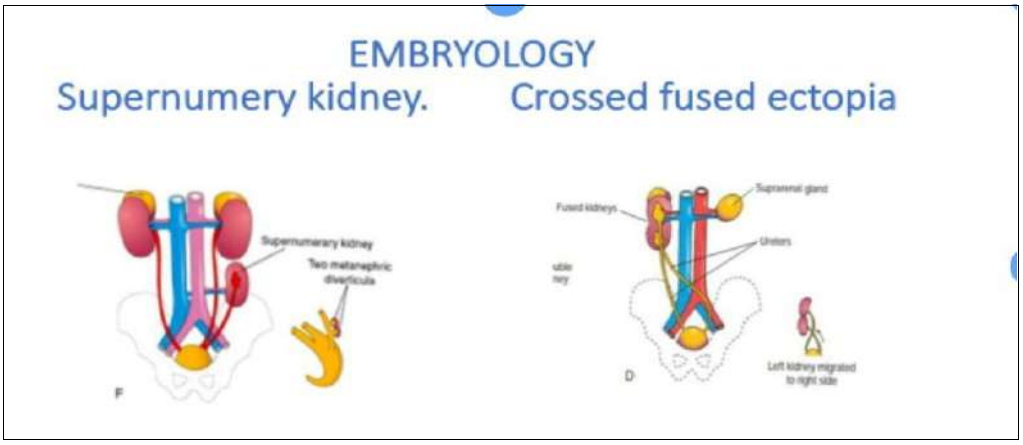
Abstract
Supernumerary kidney is the rarest of all renal anomalies; fewer than 100 cases have been reported in the literature over the years. Supernumerary kidneys are most located on the left side of the abdomen. Different pathologic conditions are reported to affect supernumerary kidneys and they may be associated with malformations of the upper urinary tract and genital tract. Because of the scarcity of reported cases and atypical presentation, they frequently causes diagnostic difficulties. Incidence of Crossed fused kidney is 1:2000, second most common renal anomaly after Horseshoe kidney. Here we report a case of 20 year old male of unilateral supernumerary kidney on left side and crossed fused ectopia on right side. Supernumery kidney with crossed fused ectopia is the rarest renal anomaly.

Keywords: eTEP, ventral abdominal hernia repair, inguinal hernias, umbilical hernia

Introduction
Supernumerary kidney is a rare congenital anomaly. It results from the aberrant division of the nephrogenic cord into two metanephricblastemas at 5th to 7th week gestation; two kidneys then form in association with a partially or completely duplicated ureteral bud. The presence of an additional ureteric bud or a branching one from the initial buds also a necessary step in embryogenesis of supernumerary kidney. Crossed fused ectopia results as a consequence of abnormal renal ascent in embryogenesis with the fusion of kidney within the pelvis around 4rth to 8th week of fetal life.



Case details
A 20 year old male patient presented with frequent attacks of acute left flank pain with difficulty in micturition and lower abdominal distension. General examination of the patient revealed thin built and tachycardia (Pulse -108/min). No other abnormality detected.
Past history revealed patient had 18-20 such episodes. No significant family history was noted.
Routine haematological and Biochemical investigations were within normal limits. Ultrasonography of the abdomen showed Left renal fossa empty. Left kidney measures 7.5x2.4 cm2 size with hilum facing anterior and fused upper pole with lower pole of right kidney suggestive of crossed fused kidney.

Approx. 13.4x9.3 cm2 sized kidney like structure noted in pelvis more towards left side with gross hydronephrosis replacing whole renal parenchyma. Ureter not visualised, possibility of PUJ obstruction in supernumerary kidney.

CT scan of abdomen without contrast showed left renal fossa empty, left kidney is noted in right lumbar region, inferior and medial to right kidney. Lower pole of right kidney is fused with upper pole of left kidney. Evidence of large approx. 11(AP) x 15(TRANS) x 18(SI)mm sized well defined fluid density cystic structure is seen in lower abdomen extending from L2 to S3 region. Approx. 5-6mm sized thickness homogenous soft tissue is noted on right lateral and posterior aspect of cystic may represent renal parenchyma. [FIG-1]. It causes displacement of adjacent bowel loops and compression over left iliac vessels. PCN was inserted in this cystic collection, which had stat output of 2000cc clear urine,

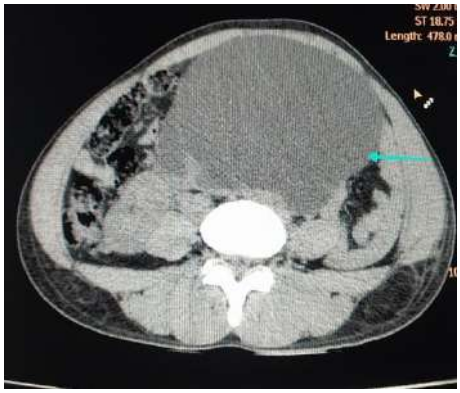


Fig 1: Axial non contrast CT showing gross hydronephrosis of supernumerary

Later, Intravenous-contrast enhanced CT depicted left renal fossa empty, left kidney noted in right lumbar region inferior and medial to right kidney. Lower pole of the rightkidney is fused with upper pole of left kidney, both showing normal cortical enhancement and prompt excretion of contrast into single pelvis and ureter into urinary bladder. Another 3rd kidney in infraumbilical region in the midline, measures 63x23mm in size, It is fused with lower pole of upper Right kidney. On administration of IV contrast, 3rd kidney shows normal cortical enhancement and prompt excretion of contrast into duplicated pelvicalyceal ureteric system, which is going into small contrast filled collection measures 14x9 mm on postero-left lateral aspect of urinary bladder. Distal ureter not visualized.

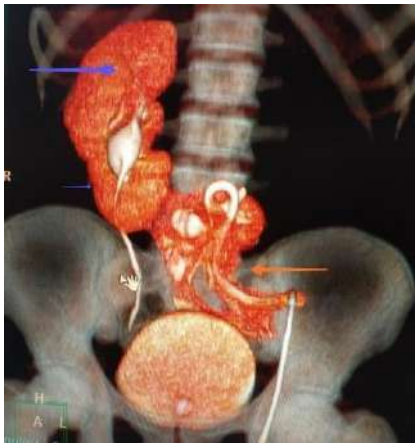


Fig 2: Coronal MIP reconstruction of contrast enhanced CT. 3D volume rendering showing supernumery kidney (orange arrow) attached to lower pole of right Crossed fused kidney (Blue arrow

Upper pole of the Left ectopia kidney is attached to the lower pole of right kidney draining into single pelvis and ureter into urinary bladder.

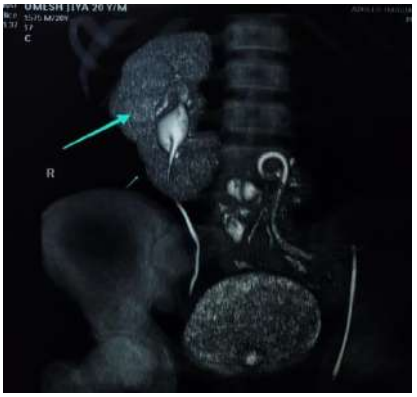


Fig 3: Coronal MI reconstruction showing the vein of Right crossed fused kidney (Blue arrow) originating from abdominal aorta and supernumerary kidney (Red arrow) has its own origin from left common iliac vein

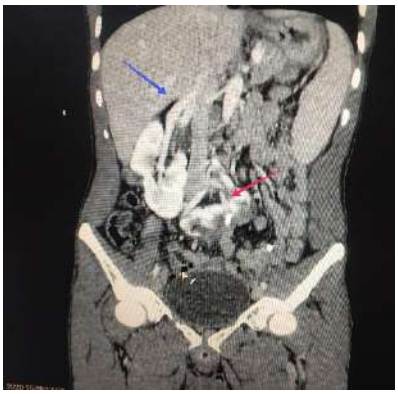


Fig 4: Coronal MIP reconstruction showing of contrast enhanced CT shows Upper pole of left kidney (Small arrow) attached to lower pole of Right kidney(Big arrow) both draining into single pelvis and ureter into urinary bladder.

Nephrostomogram was carried out in this patient by injecting 40cc iohexol dye diluted 1:1 with NS through PCN tube. On introduction of the dye pelvicalcyceal system was well outlined by dye and moderately dilated, showing complete obstruction at pelviureteric junction [FIG-5]

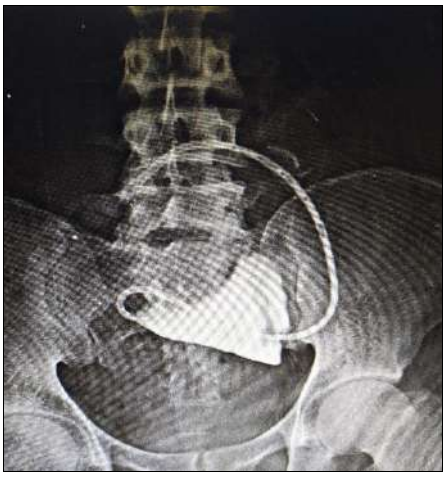


Fig 5: Nephrostogram showing complete obstruction at pelviureteric junction of supernumerary kidney**.**

Discussion

Bilateral supernumerary kidney is an uncommon renal abnormality, which influences both genders evenly [4] The supernumerary kidneys may be established in the iliac or sacral region [5]. A narrow stratum of fibrous structure may closely separate supernumerary kidneys or it is entirely discrete from the opposite side. Embryologically, the fundamental difference must discriminate the rare supernumerary kidney from the usual combined duplex systems. A sprout diverges and every division permeates a freely mixed metanephric mesenchymal mass and bifid ureters in combined duplex bifid kidneys. Once two sprouts originate disparately from the Wolffian duct and permeate the similar metanephric mass, two free nephritic collecting structures shape, but the parenchymas abide blended. A sprout divides into two and every division permeates separately a metanephric mass, which improves into unrelated simple kidneys in supernumerary kidneys with bifid ureters [6]. A supernumerary kidney may be of same size as, larger than, or more commonly smaller than the usual kidney. In most of the cases it functions normally, possess a normal shape and capsule, and is either not attached to or loosely attached to the normal kidney but in an abnormal location [3]. Most commonly they are located on the left side of the abdomen, caudal to normal kidney in our case. Coarctation of the aorta, ectopic ureteral opening, duplication of the penis or female urethra, vaginal atresia and horseshoe kidneys are congenital anomalies associated with the supernumerary kidney [7] Associated with supernumerary kidneys, many complications such as pyelonephritis, stones, pyonephrosis, hydronephrosis, and malignant changes (clear cell carcinoma, Wilms' tumor) have been reported [8]. However, the position of the extra kidney has been quite variable from case to case. It may be located in front, below, above, or behind the usual kidney. They can also be found in the iliac region or anterior to the sacral promontory.In our case the supernumerary kidney was located caudal to normal kidney and separated from the normal kidney. Supernumerary kidney is considered an accessory organ with its own arterial supply, venous drainage collecting duct and encapsulated capsule and may have separate ureter. In our case supernumerary Kidney had its own ureter. Symptoms have been noted in approximately two-thirds of the reported cases of supernumerary kidney. When symptomatic they may cause fever, pain, or palpable abdominal mass. For the diagnosis of supernumerary kidney IVP, ultrasonography, nuclear scintigraphy, CT, and MRI may be used.

Supernumerary kidney is usually smaller in size with reduced function. In our case CT IVP was diagnostic and showed Crossed fused kidney on right side and supernumerary kidney below it. Supernumerary kidney he associated with urogenital malformations like horse shoe kidney, ectopic ureteric opening, wilms tumour. In our case supernumerary kidney presented with gross hydronephrosis. With congenital PUJ obstruction. Long term follow up is needed in supernumerary kidney. In our case supernumerary kidney presented with gross hydronephrosis in which we inserted Percutaneous nephrostomy tube. Once the patient is settled we will plan for Pyeloplasty for PUJ obstruction.

Conclusion

Supernumerary kidney is a rare congenital renal malformation may be associated with different urogenital malformations. Due to high rate of complications diagnosis is very important. But due to infrequent occurrence diagnosis is challenging in many cases.

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