National Board of Examination - Journal of Medical Sciences

Volume 1, Issue 4, Pages 228–232, April 2023

DOI 10.61770/NBEJMS.2023.v01.i04.007

CASE REPORT

An Interesting case of Supernumerary Kidney with PUJ obstruction in the native kidneys

Mohammed Ayub Karamnabi Siddiqui, Ojas Vijayanand Potdar*, Vivek Shaw, Shashank Sharma, Kaustubh Vaidya and Prashant Sarawade

Grant Government Medical College and J.J. Group of Hospitals, Mumbai Central, Mumbai, Maharashtra-400008

Accepted: 12-March-2023 / Published Online: 01-April-2023

Abstract:

Congenital anomalies of Kidney and Urinary Tract (CAKUT) present with interesting challenges to a urologist in his practice. The incidence of CAKUT is 4.2 per 10,000 births. CAKUT are one of the major risk factors in adults requiring renal replacement therapy. Although children with CAKUT are often asymptomatic, CAKUT are estimated to be implicated in 30% to 60% of cases of childhood-onset chronic kidney disease (CKD) in different populations. We hereby present a rare case of a patient with Supernumerary Kidneys in which both bilateral native kidneys showed evidence of Pelvi-ureteric junction obstruction and ectopic pelvic kidney with normal function.

Keywords: Supernumerary kidney, CAKUT, PUJ obstruction, ectopic.

*Corresponding author: Ojas Vijayanand Potdar
Email: ojaspotdar@yahoo.com

Case report

A 20-year-old male patient presented with history of difficulty in passing urine per urethraly for a period of 1 month which was insidious in onset and gradually progressed and patient developed pain in bilateral flank region which was continuous, dull aching in nature and non-radiating in nature and associated fever. He was admitted and evaluated with complete blood count, renal function test and urine was sent for culture and sensitivity. He underwent ultrasound which revealed bilateral hydronephrosis with left sided renal parenchyma thinned out and right sided kidney with decreased thickness of the renal parenchyma and the findings were suggestive of bilateral Pelvis-ureteric junction (PUJ) obstruction of the two orthotopic kidneys with pyonephrosis. Hence, patient underwent bilateral percutaneous nephrostomy insertion under ultrasound guidance. Patient was managed with nephrostomy for the next 3 weeks. Daily nephrostomy output on the right side was 800 ml and on the left side it was 400 ml. After 3 weeks, left side percutaneous nephrostomy stopped draining. Patient was then evaluated using Computerised tomography Intravenous Pyelography (CT IVP). This showed evidence of two orthotopic kidneys and an ectopic right kidney located in right iliac fossa along with fusion of right orthotopic and ectopic kidney. The right sided orthotopic kidney showed gross hydronephrosis and caliectasis with thinning of right renal parenchyma with non-visualisation Left kidney showed enlarged kidney with gross hydronephrosis and thinning of renal parenchyma with transient delayed visualisation till the level of mid-ureter. The right sided ectopic kidney was located in the right iliac fossa.

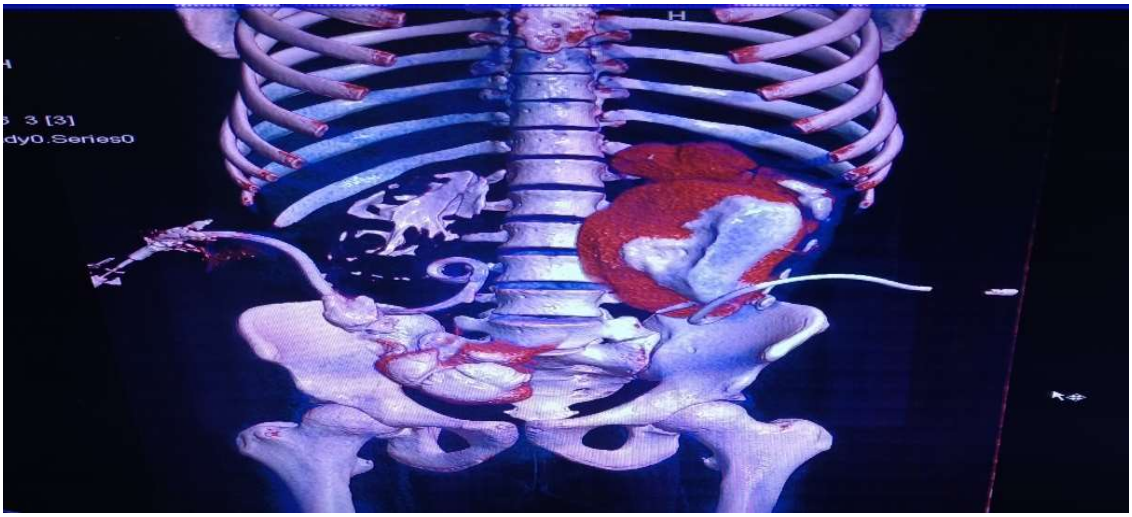

Figure 1. Reconstructed CT image shows evidence of two orthotopic kidneys with grossly dilated bilateral hydronephrosis with bilateral nephrostomy tubes in situ. Also seen is ectopic right sided kidney in the right iliac fossa.

Decision was made to internalize the drainage and to do bilateral DJ stenting of the two orthotopic kidneys. Patient then underwent Cystoscopy proceed Retrograde Pyelography (RGP) with Bilateral DJ stenting. RGP findings revealed evidence of gross hydronephrosis of right sided kidney with delayed drainage; the right sided orthotopic kidney fused with the ureter from the right sided ectopic kidney at the level of L4 level and then draining into the bladder. Left sided RGP showed evidence of gross hydronephrosis more than the right side with tapering at the level of PUJ suggestive of Left sided PUJ obstruction.

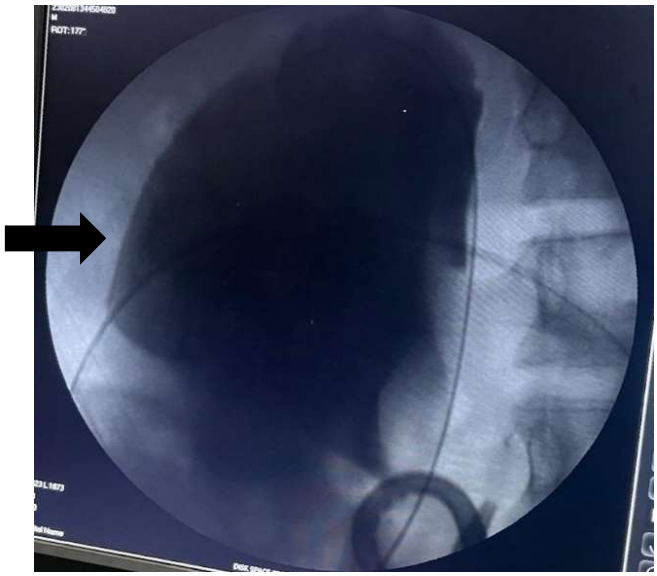

Figure 2a. Right sided RGP shows evidence of gross hydronephrosis of right orthotopic kidney with delayed drainage showing evidence of right sided PUJ obstruction.

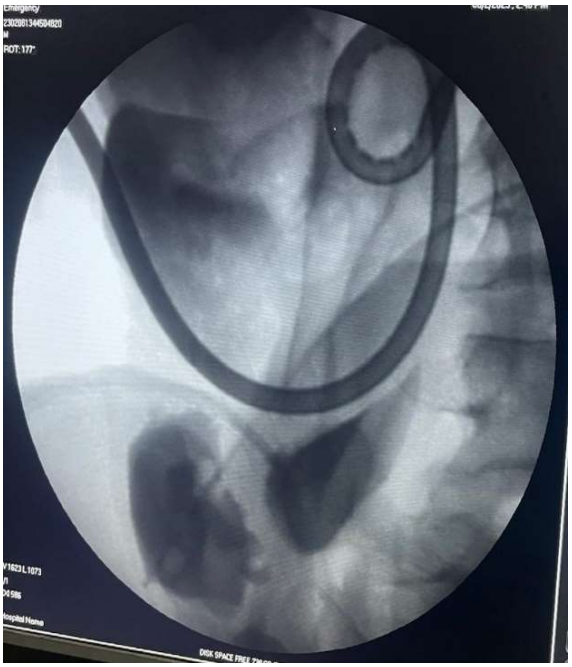

Figure 2b. Right sided RGP shows evidence of ectopic right sided kidney in the right iliac fossa with normal excretion.

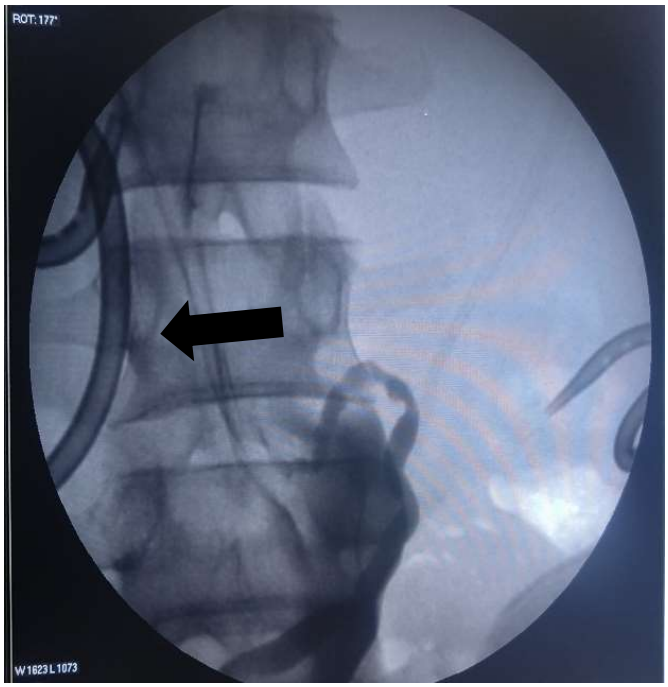

Figure 3. Left sided RGP shows evidence of kink and narrowing at the level of left sided RGP and a grossly dilated left sided renal pelvis; on account of the gross dilatation of the pelvis; contrast is not visualised in the left renal pelvis in the static images as it got diluted in the system; however, a jet going into the left sided renal pelvis was seen during the dynamic images.

Then the patient underwent DTPA scan to know the functional status of the orthotopic and the ectopic kidneys. It revealed that the orthotopic Left kidney showed adequate cortical function with obstructed excretion at PUJ level. The orthotopic and the ectopic right kidneys showed severe cortical dysfunction (<5 % combined function).

Management

The patient underwent left sided open pyeloplasty followed by open right sided nephrectomy of the native and the orthotopic right kidneys after 4 weeks. He then underwent Left sided DJ stent removal with retrograde pyelography for the left kidney 8 weeks after the pyeloplasty and it revealed prompt drainage of the left sided pelvis-collecting system with a good dependent funnel-shaped PUJ.

Discussion

Congenital anomalies of kidneys (CAKUT) presentation in patients can vary from being an incidental finding detected on imaging to a patient with renal failure or anomaly incompetent with life and stillbirth. Management of these anomalies presents a challenging task to the concerned urologist. Supernumerary Kidney may arise from irregular and abnormal division of the nephrogenic cord, which gets divided into two metanephric blastemas, which ultimately develop into two kidneys with incomplete or double ureteral bud [1-3]. Around the fifth to seventh week of gestation, when urogenital system development occurs, embryologically this anomaly used to develop. The rarity of this anomaly, its varying appearance, and the paucity of literature evidence make diagnosis and treatment difficult [4,5].

SK is an unusual urinary system congenital anomaly, with just around a hundred cases recorded in past research. It might be completely separated from the usual kidney or attached by a loose areolar tissue and it is normally smaller and less functioning as compared to normal kidneys [6]. SK is a rare congenital anomaly of the urogenital system in

which an accessory/extra or third present along with two normally located kidneys.

In our case, it was a supernumerary kidney in the right iliac fossa which was fused with the orthotopic kidney on the right side; the pelvicalyceal system of the right sided kidney showed evidence of PUJ obstruction and its ureter fused with the ureter arising from the ectopic kidney and both then draining into the bladder. The left sided orthotopic kidney also showed evidence of PUJ obstruction.

Conflicts of interest

The authors declares that they do not have conflict of interest.

References

1. Nimkar S V, Yelne P, Gaidhane S A, et al. (November 13, 2022) Supernumerary Kidney (Triple Kidney) With Horseshoe Malformation: A Case Report. Cureus 14(11): e31436. doi:10.7759/cureus.31436
2. Oto A, Kerimoğlu U, Eskiçorapçi S, Hazirolan T, Tekgül S. Bilateral supernumerary kidney: imaging findings. JBR-BTR. 2002 Dec;85(6):300-3.
3. Tada Y, Kokado Y, Hashinaka Y, Kadowaki T, Takasugi Y, Shin T, Tsukaguchi I. Free supernumerary kidney: a case report and review. J Urol. 1981 Aug;126(2):231-2. doi: 10.1016/s0022-5347(17)54457-2.
4. Mutlubas F, Mir S, Sevinc E, Türker Ö: Unilateral double kidney with contralateral supernumerary kidney which found incidentally. Ege Tıp Dergisi / Ege Journal of Medicine. 2007, 46:171-3.
5. Bernik TR, Ravnic DJ, Bernik SF, Wallack MK: Ectopic supernumerary kidney, a cause of para-aortic mass: case report and review. Am Surg. 2001, 67:657-9.
6. Ramanathan S, Kumar D, Khanna M, Al Heidous M, Sheikh A, Virmani V, Palaniappan Y. Multi-modality imaging review of congenital abnormalities of kidney and upper urinary tract. World J Radiol. 2016 Feb 28;8(2):132-41. doi: 10.4329/wjr. v8.i2.132. .