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CASE REPORT

UNILATERAL EXTRARENAL CALYCES WITH MULTIPLE CONGENITAL ANOMALIES IN AN ELDERLY MALE CADAVER

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Abstract. *The extrarenal calyces (ERCs) are very rare congenital anomalies with approximately less than 100 reported cases. ERC usually does not cause any significant problem but can be the cause of urinary stasis and infection. During routine dissection of a 69-years-old-male cadaver, three extra renal major calyces were observed at the hilum of the left kidney. We also observed an arched left testicular artery; an accessory left renal artery, double right renal veins and bilateral small renal cysts of varying sizes. ERCs often co-exist with other congenital anomalies, and such constellations of anomalies should be looked for.*

Key words: *extrarenal calyces, accessory renal artery, arched testicular artery, congenital renal anomaly*

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INTRODUCTION

Extra renal calyces (ERCs) are a very rare congenital anomaly of the urinary system wherein the major calyces, minor calyces and the renal pelvis are situated outside the parenchyma of the kidneys [1]. It was first described by Eisendrath in 1925 and later narrated in detail by Malament during the 1960s [2]. Urine produced by the nephrons of the kidney passes through the different segments of the nephron and is eventually transported and excreted out through the collecting system comprising of successive channels of the collecting tubules, collecting ducts, minor calyces, major calyces, renal pelvis, and the ureter, which empties it to the urinary bladder. Usually, there are two to three major calyces lying within the renal sinus, which empties into the renal pelvis. In ERC, however, they are found located outside the renal sinus. The accurate prevalence of ERC is not known and only less than 100 cases have been reported to date. ERC is often found associated with other congenital anomalies of the urinary system collectively referred to as the congenital anomalies of the kidneys and the urinary tracts (CAKUT); the most common association observed is that with ectopic kidney and hydronephrosis. As the condition is extremely rare and causes only insignificant, non-life-threatening symptoms, an accurate preoperative diagnosis is usually not

sought for by clinicians in most instances. However, ERC can cause prolonged stasis leading to recurrent urinary tract infection and calculi formation [3]. Furthermore, the bare calyces lying outside the kidney are also more prone for extrinsic compression by abnormal vessels and bands of tissues or for iatrogenic injury. A pre-operative diagnosis, if available, can definitely minimize the chance of iatrogenic damage to the ERC. We present a cadaveric case of left sided ERC with other coexisting congenital renal anomalies.

CLINICAL CASE DESCRIPTION

The present case of ERC was observed during routine dissection performed for undergraduate teaching in a 69-year-old male cadaver. The cadaver was received through the body donation with signed informed consent obtained from the donor's family. The institutional guidelines for using cadaveric specimens for medical teaching and research were followed. The pararenal fat was cleared, exploration and meticulous clearing of the perinephric fat within the renal fascia was performed and the renal hilar structures were exposed. On the left side, three major calyces lying outside the renal sinus draining into the renal pelvis were seen (Fig. 1b and Fig. 2). The lengths of the superior, middle and inferior ERCs on the left side were 3.4 cm, 1.9 cm and 1.4 cm, respectively. The middle and the inferior major calyces joined to form a common channel, which opened into the left renal pelvis. The superior calyx opened directly into the left renal pelvis. The left renal pelvis continued as the left ureter at the inferior pole of the left kidney and descended for 21.4 cm maintaining normal relations and opened into the respective side of the urinary bladder. We observed an arched testicular artery on the left side, which originated from the anterolateral aspect of the abdominal aorta distal to level of origin of the superior mesenteric artery in a plane posterior to the left renal vein (LRV) (Fig 1b and Fig 2). It ascended up posteriorly to the LRV reaching its superior margin. Then it arched forward, descending anteriorly to the LRV along with the left testicular vein to reach the left deep inguinal ring. An accessory LRV originating from the left lateral aspect of the abdominal aorta at L3 vertebral level, approximately at 2.3 cm below the origin of the left main renal artery, was seen. The accessory left renal artery ran for about 6.4 cm behind to the left ureter and left testicular vessels and finally supplied the anterior inferior segment of the left kidney. On the right side, two renal veins of almost equal caliber draining into the inferior vena cava were found (Fig. 1a and Fig 2). Right testicular vessels had normal courses. Multiple cystic lesions of various sizes were noted on both sides.

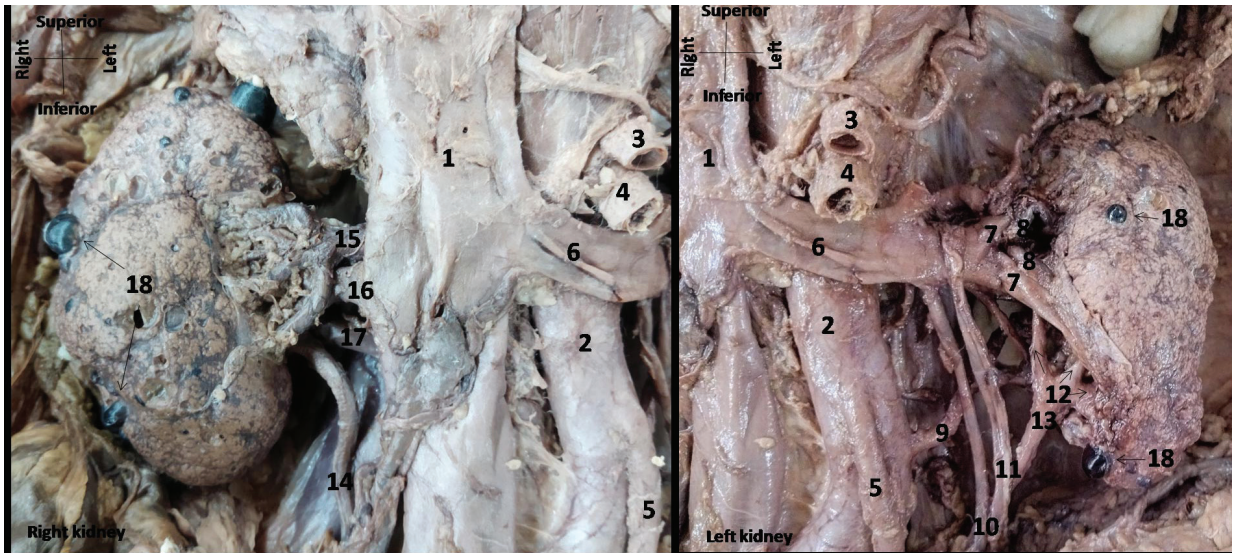

Fig. 1. Dissected kidneys lying on the posterior abdominal wall showing the various anomalies

Figure legend: 1 – inferior vena cava, 2 – abdominal aorta, 3 – celiac trunk, 4 – superior mesenteric artery, 5 – inferior mesenteric artery, 6 – left renal vein, 7 – segmental tributary of left renal vein, 8 – segmental branch of left renal artery, 9 – accessory renal artery from abdominal aorta supplying the left kidney, 10 – left testicular vein, 11 – arched left testicular artery, 12 – extra renal calyces of left kidney, 13 – left renal uretropelvic junction, 14 – right ureter, 15 – right renal artery, 16 – right renal vein, 17 – accessory right renal vein, 18 – multiple cysts

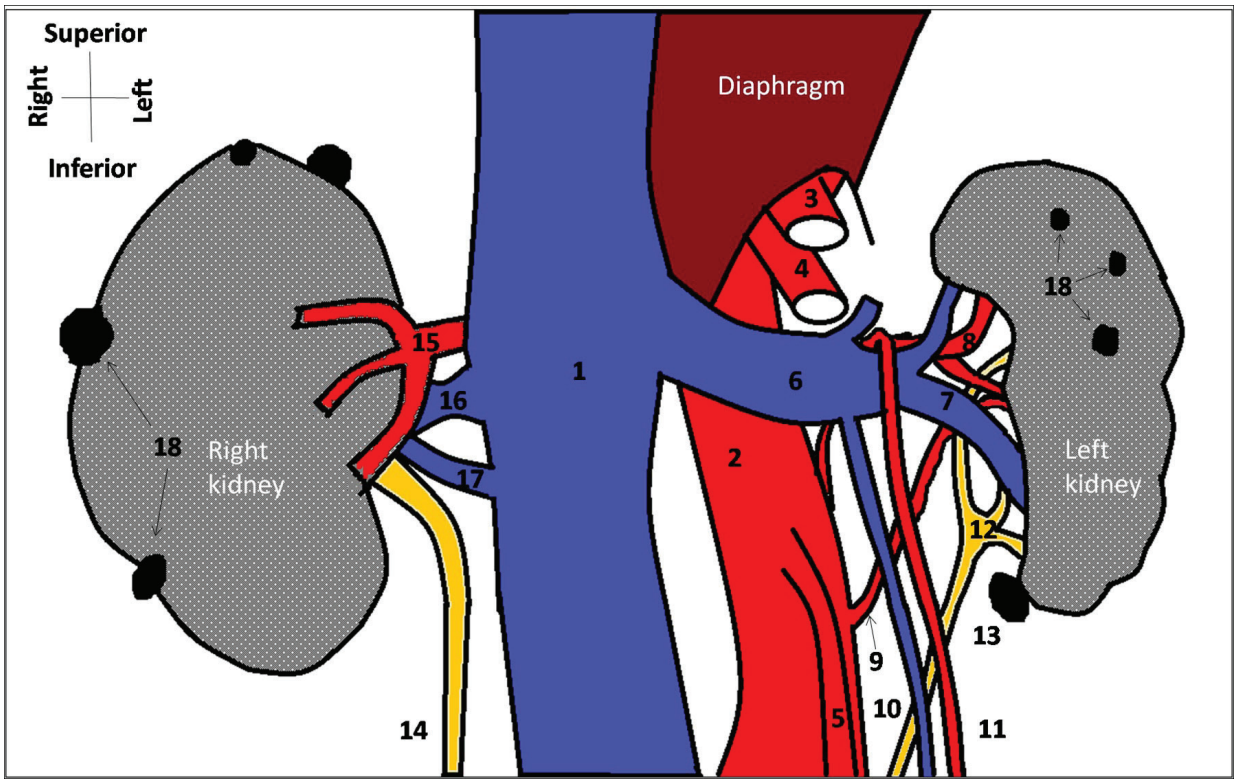

Fig. 2. Illustration depicting the morphology of the right and left kidneys and other associated vascular variations

Figure legend: 1 – inferior vena cava, 2 – abdominal aorta, 3 – celiac trunk, 4 – superior mesenteric artery, 5 – inferior mesenteric artery, 6 – left renal vein, 7 – segmental tributary of left renal vein, 8 – segmental branch of left renal artery, 9 – accessory renal artery from abdominal aorta supplying the left kidney, 10 – left testicular vein, 11 – arched left testicular artery, 12 – extra renal calyces of left kidney, 13 – left ureter, 14 – right ureter, 15 – right renal artery, 16 – right renal vein, 17 – accessory right renal vein, 18 – multiple cysts.

DISCUSSION

Exact mechanisms behind ERC causation are not known; however, faulty molecular regulation during embryonic development of the urinary system is implicated in its causation. The embryonic development of the human urinary system exhibits consecutive succession through the three nephrogenic systems viz. pronephros, mesonephros and metanehphros derived the intermediate mesoderm [4]. Malament believed it to be the result of disparity in development of ureteric bud and the metanephric blastema, resulting in failure of reciprocal interactions between the metanephric tissues with the mesonephric duct derived collecting system. As a result, more rapid development of the collecting system ensues which places the pelvicalyceal system outside the kidney [5].

Usually, ERC is an incidental finding often surprising the surgeons during autopsy or surgery. No accurate prevalence of ERC is known. A recent Indian study revealed 1% incidence of ERC in a sample of 100 autopsy specimens [6]. ERC has many reported associations with other CAKUTs, the most notable being with hydronephrosis and ectopic kidney. Hydronephrosis is often due to concomitant obstruction due to calculi. When found associated with renal calculi, ERC can complicate surgical management [7]. When associated with hydronephrosis, ERC demonstrated hydropic changes in the kidney along with ureteric wall thickening [8]. Ureteropelvic junction obstruction (UPJO) is rarely reported with ERCs. ERC with horseshoe kidney is also reported [9]. A brief account of the recently reported ERCs and the associated congenital anomalies and complications are presented in Table 1. ERCs can mimic a retroperitoneal mass or renal duplication confusing radiological diagnosis [8]. ERCs have been reported to be more common on the left side than the right side, but no suitable explanation for that is known [10]. Clinical presentation can vary depending on the association with other CAKUT. It can be a simple presentation of urinary tract infection or with symptoms due to associated CAKUT such as a loin mass, hydronephrosis, etc. ERC with PUJ obstruction, hydronephrosis, calculi, etc., usually requires surgical correction of ERC. Intravenous urography and magnetic resonance urogram can

Table 1. Reports of extrarenal calyces associated with other congenital anomalies in the last two decades

Authors	Population/sample size	Study type	Findings
Garg P et al., 2002 [3]	Indian, single case	Radiological	Both minor and major calyces located outside the kidney
Taha S A et al., 2006 [11]	Saudi, single case	Surgical	ERC with renal dysplasia in a 2-year-old male child
Nataraju G et al., 2009 [12]	Indian, single case	Surgical	Atypical ERC and hydronephrosis associated with ureteric atresia in a 12-year-old male child
Raghunath B. V et al., 2012 [13]	Indian, single case	Surgical	ERC associated with pelviureteric junction obstruction
Gupta T et al., 2014 [5]	Indian, two cases	Autopsy	2 cases of ERC wherein calyces and renal pelvis are located outside the kidney
Gandhi K R et al., 2015 [14]	Indian, single case	Autopsy	ERC with multiple stones in the
Suwannakhan A et al., 2016 [15]	Thai, single case	Autopsy	Multiple renal anomalies associated with ERC
Sahni D et al., 2017 [16]	Indian, 1900 cases	Autopsy	ERC was associated with renal agenesis, renal ectopia with fusion, trilobar kidney, polycystic kidney.
Rajendran S et al., 2017 [1]	British, single case	Surgical	Multicystic dysplastic kidney giving in association with six extra renal major calyces.
Maldonado A A G et al., 2018 [10]	Mexican, single case	Surgical	ERC presented as severe hydronephrosis in a 6-year-old female child.
Wahyudi I et al., 2019 [2]	Indonesian, single case	Surgical	ERC mimicking retroperitoneal mass.
Ahuja S et al., 2021[17]	American, single case	Autopsy	ERC associated with renal vascular variation
Khanna V et al., 2022 [8]	Indian, single case	Surgical	ERC with pelviureteric junction obstruction in a 10-year-old male child

readily delineate ERC and are also helpful for looking at coexisting CAKUT. They can also provide an idea about the functional status of the kidney. Early in the 1960s. Malament et al. described “bowing outwards of calyces with terminal incurvation” resembling a “small hand garden rake” as the diagnostic radiological sign. The preferred choice of the surgical procedure in complicated symptomatic ERC is dismembered pyeloplasty. Very recently, a unique method of calyx unification and subsequent anastomosis to the ureter has been demonstrated to be successful [2]. As with most ERCs, the present case also remained asymptomatic throughout lifetime, in spite of harboring ERC with associated congenital renal anomalies. This indicates that the prevalence of ERCs can be much higher than expected.

The reported incidence of ‘arched testicular artery’, which is also referred to as the ‘arched gonadal artery of Luschka’, is around 6%. The arched left testicular artery observed here, which is similar to that observed by Naito et al. in two cases, can be classified as type-III variety of the gonadal arteries as per Notkovich’s classification system. The arching over the LRV is particularly important as it can cause compression of the ipsilateral renal vein impeding the venous return not only from the left kidney, but also from the left testis. Consequently varicocele, orthostatic proteinuria and hematuria can occur. Furthermore, the possibility of an unusual nutcracker phenomenon like condition arising out of the LRV compression by the left testicular artery also cannot be ruled out. The other variation observed on the left side was accessory renal artery, which is a relatively frequent variation with an incidence of up to 30%. Accessory renal artery is more frequently reported on the right side. Additionally, multiple solid lesions most probably a sequel of cystic lesions were seen on both kidneys with double renal vein draining the right kidney. The present case exhibits a rare situation of extra renal calyces associated with multiple congenital anomalies of the kidneys and visceral vessels around it. In the present case, it remained quiescent without apparently causing any clinical manifestation throughout life. In this particular case, ERC on the left side was coexisting with variations of multiple visceral vessels which hold special importance, as insufficient knowledge about them may prove critical during surgical management for many conditions such as renal transplant, varicocele, renal or ureteric stone, etc. It can cause diagnostic confusions by mimicking as a retroperitoneal cyst in imaging studies. Considering its association with congenital anomalies, a clinician dealing with any CAKUT or other renal vascular variation should be vigilant for possible presence of ERC.

CONCLUSIONS

The concurrent association of extra renal calyces with multiple congenital malformations of the kidney is a rare scenario. Existing evidence suggests that

extra renal calyces can be actually more prevalent than believed. Given the frequent association with other congenital anomalies in most of the reported cases, including the present case, it is reiterated that ERC should be suspected while dealing with any other congenital anomaly of the kidney and urinary system. The coexistence of multiple anomalies further point towards the faulty regulation of the embryonic development by some common factors involved in the embryonic regulation. Clinicians should be aware of the possibility of encountering the ERC while dealing with other CAKUTs, as it would prevent diagnostic confusion and unwanted injury to the pelvicalyceal system.

Abbreviations used:

CAKUT: *congenital anomalies of the kidneys and the urinary tracts*
ERC: *extrarenal calyces*
LRV: *left renal vein*
UPJO: *Ureteropelvic junction obstruction*

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Authors' Contributions:

D. Borthakur – Protocol development, reviewed literature, cadaveric dissection, manuscript writing.
R. Kumar – Protocol development, reviewed literature, photography, manuscript writing.
S. Kamalesh – Reviewed literature, photography, manuscript writing.
R. Dada – Protocol development, reviewed literature, manuscript writing.

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REFERENCES

1. Rajendran S, Cho A, Mishra P, Cherian A. Hydronephrotic kidney with multiple extra-renal calyces. Ann R Coll Surg Engl. 2017;99(8):e219–20.
2. Wahyudi I, Rodjani A, Situmorang GR, et al. Extrarenal calyces mimicking retroperitoneal cystic mass with concomitant ureteropelvic junction obstruction: Renal pelvis reconstruction using calyx unification. Res Reports Urol. 2019;11:143–8.
3. Garg P, Godara R, Karwasra RK. Extrarenal calyces: A rarity. Urol Int. 2003;71(3):331–2.
4. Kozlov VM, Schedl A. Duplex kidney formation: developmental mechanisms and genetic predisposition. F1000Research. 2020;9.
5. Gupta T, Goyal SK, Aggarwal A, et al. Extrarenal calyces: a rare renal congenital anomaly. Surg Radiol Anat. 2015;37(4):407–10.
6. Anjana TSR, Muthian E, Thiagarajan S, Shanmugam S. Gross morphological study of the renal pelvicalyceal patterns in human cadaveric kidneys. Indian J Urol. 2017;33(1):36–40.
7. Gandhi KR, Chavan S. Revisiting the morphology of pelvicalyceal system in human cadaveric kidneys with a systematic review of literature. Asian J Urol. 2019;6(3):249–55.
8. Khanna V, Khanna K, Chauhan A, Kapoor R. Extrarenal calyces with ureteropelvic junction type obstruction. BMJ Case Rep. 2022;15(3):2021–2.
9. Mohanty C, Ray B, Samaratunga U SG. Horseshoe Kidney with Extrarenal Calyces – A Case Report. J Anat Soc India. 2002;51(1):57–8.
10. González Maldonado AA, Manzo Pérez G, Vanzzini Guerrero MA, et al. Extrarenal calyces as a cause of non-functional kidney in a child: Case report. Urol Case Reports. 2018;17:56–8.
11. Taha SA, Hashish MH, Eldarawany HM, et al. Renal dysplasia with extrarenal calyces. Saudi Med J. 2006;27(3):392–4.
12. Nataraju G, Nandeesh BN, Gayathri MN. Extrarenal calyces: A rare anomaly of the renal collecting system. Indian J Pathol Microbiol. 2009;52(3):368–9.
13. Raghunath B V, Babu NM, Gowrishankar B, Ramesh S. Extrarenal calyces associated with pelviureteric junction obstruction: A case report of a rare anomaly. J Indian Assoc Pediatr Surg. 2012;17(3):124–5.
14. Gandhi KR, Wabale RN. A rare presentation of the extra renal pelvis containing multiple calculi. Pravara Med Rev. 2015;7(2):30–2.
15. Suwannakhan A, Meemon K. Horseshoe kidney with extrarenal calyces and malformed renal vessels. Eur J Anat. 2016;20(4):355–9.
16. Sahni D, Aggarwal A, Gupta T, et al. Congenital Renal Anomalies in Indian Population. J Postgrad Med Educ Res. 2017;51(1):12–6.
17. Ahuja S, Maglasang N, Tan Y, Daly DT. A unique case of extrarenal calyces and associated vascular variations in an adult female cadaver. Folia Morphol. 2022;81(4):1072–8.