Extrarenal calyces in a pelvic kidney with ureteropelvic junction obstruction in an adult male – A rare case report

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Abstract Extrarenal calyces (ERC) is a rare renal anomaly. First described in 1925, and till now, >60 cases have been reported worldwide. The association of ERC in ectopic kidneys with ureteropelvic junction obstruction (UPJO) is a very rare presentation. We encountered a case of an adult male with ERC in a pelvic kidney with UPJO, in which the dilated ERC mimicked the ureter and created intraoperative confusion.

Keywords: Extrarenal calyces, pelvic kidney, ureteropelvic junction obstruction

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INTRODUCTION

Extrarenal calyces (ERC) is a rare renal anomaly and can present as flank pain. ERC can have complex radiological findings or can cause intraoperative confusion. It was first described in 1925, and <60 cases are reported in literature worldwide.^[1] These ERC lie outside the renal parenchyma and are connected to the pelvis. They are seen as multiple tubes (3–5 in number) coming out of the renal hilum. It is often associated with other renal anomalies like hydronephrosis due to ureteropelvic junction obstruction (UPJO) and renal dysplasia.^[2] The association of ERC in ectopic kidneys with UPJO is a very rare presentation. To our knowledge, two cases of ERC in ectopic kidneys with UPJO have been reported in pediatric patients.^[3] We present a report of an adult male with ERC in a pelvic kidney with UPJO.

CASE REPORT

A young male presented with complaints of left flank

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pain for the past 4 months. On evaluation with computed tomography (CT), urography left kidney was found to be malrotated with renal hilum facing anteroinferiorly and ectopically in the pelvic region at L3-S1 vertebral level. CT also revealed mild dilatation of the calyceal system with significant ballooning of renal pelvis (AP diameter 87 mm) indenting on the dome of UB with abrupt tapering at pelviureteric junction (PUJ) with the collapse of the distal ureter and nonopacification on a delayed scan [Figure 1]. He was further evaluated with diethylenetriaminepentaacetic acid (DTPA) (99 mTc DTPA diuretic renal scan) suggesting ectopically located grossly hydronephrotic left kidney with impaired function (35%) and delayed clearance. He was taken up for robot-assisted left pyeloplasty. Intraoperatively, a retrograde pyelogram was done, suggesting nondilated ureter with spillage of contrast into the dilated pelvis with acute lateral angulation of PUJ at L5-S1 level [Figure 2]. Endoscopically, a grossly hydronephrotic pelvic kidney was seen. The dilated and convoluted ERC mimicked

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the ureter [Figure 3]. In total, 3 ERC were identified intraoperatively. On meticulous dissection, the ureter was traced retrogradely from the vesical end [Figure 4]. PUJ was identified, and dismembered Anderson–Hyne's pyeloplasty was done to repair the UPJO with double J stent across the anastomosis. The patient was discharged in stable condition on postoperative day 5. His follow-up scans after 6 weeks of DJ stent removal showed nonobstructive clearance and adequate function.

DISCUSSION

UPJO is present in 20%–40% of cases of the ectopic kidney. Renal ectopia is usually associated with malrotation with possible vascular aberrations. UPJO in a pelvic kidney with ERC is a much rarer association. Defining surgical anatomy is necessary for managing such aberrations. Prediction of ERC in a preoperative setting by CT is difficult, and it



Figure 1: The CT urography findings of the pelvic kidney with UPJO in the axial and coronal section. UPJO: Ureteropelvic junction obstruction, CT: Computed tomography



Figure 3: Intraoperative finding of ERC (3 in number, denoted by arrows). ERC: Extrarenal calyces

usually presents as a surprise finding intraoperatively. We also encountered a case in which the dilated and elongated ERC mimicked the ureteral diverticulum.

The exact cause of ERC is not very clear. One of the accepted hypotheses suggests the disparity between the rate of development of metanephric tissue and ureteric bud as a potential cause of ERC. If the ureteric bud has a rapid development, the calyceal system could well develop before its coalescence with the nephrogenic mass. Conversely, a delay in the growth of the nephrogenic mass delays its attachment to the collecting system, leading to extrarenal development of the collecting system.^[4,5]

Treatment of this condition is based on the functional status of the involved kidney. Pyeloplasty should be considered in the kidneys with preserved renal function.



Figure 2: Retrograde pyelogram showing acute angulation of ureter near UPJ and filling of the pelvis with the contrast (long arrow denotes the UPJ; short arrow denotes the dilated pelvis). UPJ: Ureteropelvic junction



Figure 4: Intraoperative image of ureter and UPJ, the figure below shows the ureteral catheter *in situ*. UPJ: Ureteropelvic junction

Pelvis reduction or resection is not required. While doing reduction pyeloplasty, one has to keep in mind the possibility of ligating the calyx, which may result in hydrocalyx.

ERC in pelvic kidneys with UPJO has been a rare entity. To our knowledge, two such cases have been reported by Kanojia *et al.*^[3] In their case report, both patients were of the pediatric age group; however, we encountered UPJO with ERC in a pelvic kidney in an adult male.

CONCLUSION

ERC with PUJO in a pelvic kidney is a very rare condition, and urologists should be aware of this anatomical entity. ERC is encountered intraoperatively as a surprise finding. Hence, the abnormal anatomy should always be defined by preoperative imaging and intraoperative retrograde pyelogram with meticulous and careful dissection.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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