Extrarenal calyces with ureteropelvic junction type obstruction

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DESCRIPTION

Extrarenal calyces (ERC) is a rare renal anomaly and can present with a solitary clinical symptom of flank pain, or as a complex radiological finding or a confusing intraoperative finding. Esiendrath first described the anomaly in 1925 and less than 50 cases are reported in literature worldwide.¹ In this anomaly the major calyces and pelvis lie outside the kidney and it is often associated with other renal anomalies like hydronephrosis due to ureteropelvic junction obstruction (UPJO), dysplasia and ectopic kidney.²

Herein, we present a case of ERC in a 10-year-old boy presenting with pain and a lump in the left flank. Ultrasound showed a hydronephrotic left kidney, size 11.3×5.1 cm with thinned out cortex and dilated pelvis likely due to UPJO. Right kidney was normal. The renal dynamic scan confirmed severe hydronephrosis and poor function (9%) in the left kidney. Open left pyeloplasty was planned for the patient. Consent for left nephroureterectomy was also taken in view of poor function. No other imaging was performed.

An anterior flank extraperitoneal approach was used to access the left kidney. Intraoperatively, the ballooned kidney was decompressed and

Figure 2 (A) Microscopic sections (10X) from kidney showing hydropic changes and (B) section from ureter showing thickened muscular wall.

150 mL of urine was drained. Abnormal intraoperative findings in form of multiple tubular structures (1–3 cm in length) arising from the kidney and then draining into a dilated sac with non-identification of the ureter during initial dissection pointed towards a developmental anomaly of the pelvicalyceal system.

On initial examination, it seemed like a variant of renal duplication anomaly. On further dissection, four major calyces were clearly identified which emerged from the kidney draining into the dilated pelvis and the ureter was attached to the lowermost part of renal pelvis and narrowing at UPJ was present.

In view of poor renal function (9%), papery thin parenchyma and complex anatomy, left nephroureterectomy was performed (figure 1). The histopathological finding showed a markedly dilated pelvicalyceal system with thickened ureter and focal hydropic changes in the kidney (figure 2). In the case with the poor function (<10%) but a good robust parenchyma and comparable renal size, we would have done pyeloplasty.

The disparity in the rate of growth that is, rapid branching of ureteric bud into calyces before it coalesces with the metanephric blastema or the delayed differentiation of the metanephric tissue has been postulated as the cause for the origin of

Figure 1 Nephroureterectomy specimen of left kidney showing papery thin parenchyma and four major calyces draining into the dilated pelvis, the extrarenal calyces (ERC).

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Learning points

- ▶ Multiple tubular calyces which lay outside the kidney should alert the surgeon towards the possibility of extrarenal calyces (ERC).
- ▶ ERC anatomy can be confused with renal duplication anomalies.
- ▶ ERC may be associated with ureteropelvic junction obstruction type of hydronephrosis.

ERC anomaly.³ The child is in our follow-up for 13 months and has been symptom free.

In such cases, if there is preoperative suspicion of ERC, an intravenous pyelography or an MR urogram can definitely prove helpful to delineate the complex anatomy and functional status of the renal moiety beforehand.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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