Hydronephrotic kidney with multiple extra-renal calyces

S Rajendran, A Cho, P Mishra, A Cherian

Urology, Great Ormond Street Hospital, London, UK

ABSTRACT

Extra-renal calyces are a rare anomaly of the renal collecting system, characterised by the presence of calyces and pelvis outside the renal parenchyma. It may also be associated with other anomalies, such as renal ectopia, fusion and malrotation. We describe an unusual case of extra-renal calyces with six long calyces arising from a suspected multicystic dysplastic kidney that was successfully managed laparoscopically.

KEYWORDS

Extra-renal calyces – Hydronephrosis – Laparoscopic nephrectomy

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CORRESPONDENCE TO

Simon Rajendran, E: simonrajendran@gmail.com

Case history

At 20 weeks of gestational age, routine prenatal ultrasound revealed the appearance of a large cystic structure in the region of the left kidney in the female fetus. Postnatal imaging showed a significantly enlarged left kidney measuring 9 cm in length with large dilated pelvis and multiple dilated calyces around it, with associated hydroureter. A dimercaptosuccinic acid renal scan performed at 2 months of age demonstrated a non-functioning left and normal functioning right kidney. A micturating cystourethrogram showed a collapsed ureterocele on the left side of the bladder with no evidence of reflux. In view of the non-functioning large left kidney, laparoscopic transperitoneal nephrectomy was planned. At laparoscopy, a left kidney with six major calyces and pelvis lying outside the renal parenchyma was identified (Fig 1). The kidney was supplied by a single renal artery and vein. Renal pelvis and major calyces were carefully dissected out and nephrectomy was performed (Fig 2). Postoperative recovery was uneventful, and the patient was discharged on the third postoperative day. Histopathology of specimen showed normal tubular structures with absence of muscle cells, features which are pathognomonic of extra-renal calyces (ERC).

Discussion

ERC, one of the rarest anomalies of the urinary collecting system, was first described by Eisendrath in 1925 and later in more detail by Malament in 1961.^{1,2} It is defined as an anomaly in which the renal pelvis, major calyces and occasionally minor calyces lie outside the renal parenchyma.³ ERC has previously been associated with other renal

anomalies such as renal ectopia, horseshoe kidney, bifid ureter and renal dysgenesis.^{2,4} The exact mechanism involved in the development of ERC is not clear. Malament proposed that ERC was due to developmental disparity resulting from slow development of metanephric tissue and relatively rapid ureteral bud development.^{1,2} The slow development of the metanephric tissue delays attachment to the collecting system, resulting in first- and second-order collecting system lying outside the renal parenchyma.² In addition, the rapid development of the ureteric bud gives rise to a mature calyceal system prior to its interaction with the nephrogenic mass.² Failure of the ureteric bud to

Figure 1 Laparoscopic view of kidney with extra-renal calyces

Figure 2 Left nephroureterectomy specimen with six extra-renal calyces

Figure 3 Ultrasound demonstrating distortion and laterally placed calyces suggesting extra-renal anatomy

effectively coalesce with the nephrogenic mass could account for the association of ERC with renal ectopia and non-rotation, contributing to inadequate stimulus for rotation and ascent.^{2,4}

In the majority of cases reported in the literature, radiological investigations failed to successfully define this anomaly and diagnosis was made intraoperatively.⁵ The rarity of

this anomaly and the ballooning effect of easily distensible unsupported ERC mimic the appearance of a dilated pelvis.² In our patient, ultrasound showed a large dilated pelvis with surrounding multiple dilated calyces, which was initially believed to be multicystic dysplastic in nature. However, in retrospective review, ERC features were appreciated (Fig 3).

ERC have also been associated with renal pelvic outflow obstruction leading to irreversible renal damage.^{6,7} The distensibility of the unsupported ERC and pelvis in contrast to a normal kidney will result in an increase in calyceal volume. It is proposed that the unsupported dilatation makes it more prone to kinking.²

Management options in patients with ERC with pelviureteric junction obstruction vary depending on renal function. In kidneys with preserved function, reconstruction by either classic pyeloplasty or ureterocalicostomy with fusion of calyces has been successful.⁸ To avoid injury in such situations, the pelvis should be opened anteromedially and the interior of the pelvis should be inspected prior to renal pelvis transection. In patients with poor function, nephroureterectomy should be performed.^{9,10} Pathological features to support distinction between ERC and ureteric duplications are the absence of muscle staining in the former.

This case highlights the importance of considering this condition where there is inconclusive imaging to aid early diagnosis and prevent further nephron damage and to help avoid inadvertent injury to pelvicalyceal systems during surgery in patients with preserved renal function.

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