CASE REPORT

Double trouble: pelvi-ureteric junction obstruction and renal cell carcinoma in right to left crossed fused ectopia

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SUMMARY

Crossed renal ectopia is a rare occurrence. The majority of the crossed units are fused to their ipsilateral mate. Pelvi-ureteric junction obstruction (PUJO) in crossed fused moieties has been reported very rarely. Similarly, malignancy arising in the crossed over moieties is a rare occurrence too. Only a few cases have been reported earlier. We are here reporting an unusual case with coexistent PUJO and renal cell carcinoma in the crossed over moiety of right to left crossed fused ectopia. To the best of our knowledge, this is the first reported case with both these pathologies, coexisting, in the crossed over moiety. The patient underwent open radical nephrectomy of the right moiety via a lower midline incision. Surgery in this scenario is very challenging due to complex anatomy with aberrant vasculature, which needs for dissection within the renal parenchyma, similar to nephron-sparing surgery to separate from the normal moiety.

BACKGROUND

Rarest of the rare scenario with coexistent pelvi-ureteric junction obstruction (PUJO) and renal cell carcinoma in fused anomalies.

CASE PRESENTATION

A man aged 35 years, with no known comorbidities, presented with a history of colicky abdominal pain in the periumbilical and suprapubic region associated with nausea, of 5 years duration. He did not have any associated urinary or bowel symptoms. Abdominal examination revealed a palpable lump in the lower abdomen. Genitalia and digital rectal examination did not reveal any abnormality.

INVESTIGATIONS

His haemogram and serum chemistries were within normal limits. Chest X-ray was normal and on abdominal ultrasonography, grossly hydronephrotic right kidney was seen in the umbilical region. On contrast-enhanced CT scan, he was found to have right to left crossed fused ectopia with PUJO in the crossed over moiety along with a 3.9×3.3 cm enhancing renal parenchymal tumour (figure 1). Renal dynamic scan revealed the right crossed over moiety to be poorly functioning (10%).

DIFFERENTIAL DIAGNOSIS

1. PUJO in an ectopic kidney
2. Tumour in an ectopic kidney
3. Retroperitoneal tumour

TREATMENT

Retrograde pyelogram revealed that the right ureter was crossing the midline towards the left with a 'Jet Sign' into the grossly hydronephrotic right crossed over moiety, suggestive of PUJO (figure 2). Ureteral access catheter was placed in the left ureter, for intraoperative delineation and prevention from inadvertent injury. He underwent open radical nephrectomy of the right moiety via a lower midline incision. Intraoperatively, the right moiety was grossly hydronephrotic and was fused with the lower pole of left moiety, lying in the midline extending into the true pelvis. The parenchyma was thinned out. There was a solid renal mass, which was located posteriorly within the right moiety (figure 3). Intraoperative blood loss was minimal.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful. Histopathology of the tumour showed features of clear cell variety of renal cell carcinoma, Fuhrman grade 2, without any capsular infiltration. At 2 months follow-up, the patient is doing fine with significant relief of his symptoms.

DISCUSSION

Crossed renal ectopia is a rare congenital anomaly and in around 90% of cases, presents with fusion of both kidneys. The autopsy incidence is reported as 1 in 2000.¹ With a slight male predominance (3:2), left to right crossover occurs more frequently than its reverse.¹ The most common anomaly involves fusion between the lower pole of the orthotopic kidney and the upper pole of the ectopic kidney. The ureter of the crossed over moiety, however, crosses the midline to insert into its orthotopic location within the bladder trigone. In our case, it was right to left crossover with two ureters inserting into their normal positions with well-developed trigone.

Barring a few case reports, PUJO in fusion anomalies has been reported very rarely. Only a few case reports are available in the published literature,²⁻³ but no significant data are available on its incidence, natural history and the management concerns of PUJO in this anatomically abnormal scenario.

The association of malignant tumours with fusion anomalies is an extremely rare event and only a few cases have been reported so far.⁴⁻⁹ Except for one case of transitional cell carcinoma,¹⁰ all other cases had renal cell carcinoma. In any type

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Figure 1 Contrast-enhanced CT abdomen: coronal (left) and axial (right); showing the right crossed over moiety fused with the lower pole of left moiety, also showing the renal mass.

Figure 2 Right retrograde pyelogram (RGP)—orthotopic location of the right orifice with crossing over of the right ureter to left across the midline. Left RGP—left ureter apposed to the grossly enlarged hydronephrotic, ectopic right moiety.

Figure 3 Cut section of the specimen.

of ectopia and fusion, the vascular supply of the orthotopic and the ectopic moiety is highly variable and unpredictable. One or more renal arteries arising at a variable level from the aorta or common iliac vessels may supply the renal units. Surgery in this scenario of fused ectopic kidneys is a challenging task due to its complex anatomy and aberrant vascular supply. This needs for dissection within the renal parenchyma, similar to nephron-sparing surgery. In our case, the crossed over fused right moiety had anomalous blood supply from the left common iliac vessels, though the main arterial branch was seen to be originating at the level of bifurcation of aorta, ventrally.

A single case report is available in the published literature with simultaneous occurrence of ureteropelvic junction obstruction and renal cell carcinoma in an orthotopic kidney.¹¹ No case has been reported so far with both the pathologies, coexisting in the ectopic kidneys.

Learning points

- ▶ Adequate preoperative work-up should be performed to delineate anomalous vascular supply.
- ▶ Renal dynamic scan should be performed to assess differential function and drainage patterns of both the moieties.
- ▶ Preoperative ureteral stenting of normal moiety should be performed in order to avoid inadvertent ureteral injury.
- ▶ Anteriorly based, trans-peritoneal approach provides for the optimal exposure of the anomalous anatomy which allows for early vascular control and improved chances for nephron sparing.

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REFERENCES

- Shapiro E, Bauer SB, Chow JS. Anomalies of the upper urinary tract. In: Wein AJ, Kavoussi LR, Novick AC, *et al.* eds. *Campbell-Walsh urology*. 10th edn. Philadelphia: Saunders, 2012:3140–5.
- Modi P, Goel R, Dodia S. Case report; Laparoscopic pyeloplasty with pyelolithotomy in crossed fused ectopia. *J Endourol* 2006;20:191–3.
- Muruganandham K, Kumar A, Kumar S. Laparoscopic pyeloplasty for ureteropelvic junction obstruction in crossed fused ectopic pelvic kidney. *Korean J Urol* 2014;55:764–7.
- Stanley KE, Winfield HN, Donovan JF, *et al.* Laparoscopic nephrectomy in crossed fused renal ectopia. *Urology* 1993;42:375–8.
- Bolton DM, Bowsher WG, Costello AJ. Renal cell carcinoma in both moieties of crossed fused ectopia. *Aust N Z J Surg* 1993;63:662–3.
- McDonald JH, McClellan DS. Crossed renal ectopia. *Am J Surg* 1957;93:995–1002.
- Sugita S, Kawashima H, Nakatani T, *et al.* Renal cell carcinoma in an L-shaped kidney. *Int J Urol* 2000;7:236–8.
- Stimac G, Dimanovski J, Ruzic B, *et al.* Tumors in kidney fusion anomalies—report of five cases and review of the literature. *Scand J Urol Nephrol* 2004;38:485–9.
- Davis CM, Rao MV, Flanigan RC, *et al.* Renal cell carcinoma in two patients with crossed fused ectopic kidneys. *Urol Int* 2008;81:370–2.
- Gur U, Yossepowitch O, Baniel J. Transitional cell carcinoma in a fused crossed ectopic kidney. *Urology* 2003;62:748.
- Abe T, Nakayama J, Kishikawa H, *et al.* Renal cell carcinoma with hydronephrosis due to pelviureteric junction obstruction: a case report. *Hinyokika Kyo* 2005;51:109–12.

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