CASE REPORT

Rare case with plethora of upper urinary tract anomalies associated with pelviureteric junction obstruction: a surgical challenge managed with robot assistance

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Accepted 23 December 2018

SUMMARY

The genitourinary system for reasons unknown is more likely to have birth defects than any other system. The anomaly of collecting system draining the kidney represent mystifying subset of congenital anomalies. Pelviureteric junction obstruction (PUJO) is most common. Chronic obstruction can lead to stasis, urinary infection and stone formation in PUJO. Extrarenal calyces, which is characterised by presence of calyces and renal pelvis outside the renal parenchyma is one of the rare anomalies seen among the collecting system right gonadal vein drains into inferior vena cava. Its altered drainage into right renal vein is rarely seen and reported. Glut of these multiple anomalies in a single case is an extremely rare event. We hereby discuss a case of 40-year-old male patient with combination of all these anomalies and discuss the embryology, presentation and management.

BACKGROUND

The genitourinary system for reasons unknown has the distinction of having most probability of congenital anomalies. Approximately 30% of the children born with congenital anomalies tend to have genitourinary anomalies. Thorough insight of these congenital aberrations may aid as a valuable

Figure 2 (A,B) Intraoperative image showing right gonadal vessels and dissection towards renal hilum. (C) Right gonadal vein draining into a branch of renal vein. (D,F) Renal pelvis dissection and excision of pelviureteric junction with pyeloplasty reconstruction over 4.8 French double J stent.

guide for urologists and can circumvent diagnostic inaccuracies, impact surgical techniques and prevent surgical complications. Advances in the imaging techniques have made identification and appropriate preoperative planning of such patients. In this paper, we present our experience of managing a patient with multiple upper urinary tract anomalies followed by brief discussion.

CASE PRESENTATION

A 40-year-old man presented with episodic colicky bilateral flank pain of 2-year duration. Pain subsided on taking oral analgesics. Patient denied any history of fever, haematuria or dysuria. There was no significant medical history.

INVESTIGATIONS

The clinical examination of the patient was unremarkable. Urinalysis showed pH of 6.1, four to six pus cells and culture was sterile. Routine lab investigations such as haemogram, kidney function test and liver function tests were normal. Ultrasound of abdomen showed bilateral hydronephrosis with bilateral nephrolithiasis. Plain X-ray kidney, ureter and urinary bladder showed right multiple round radio opaque shadows at the level of L1 vertebra and single large radio opaque shadow on left side suggestive of bilateral renal stones (figure 1A). Subsequently, an intravenous pyelography revealed right sided grade II and left sided grade

Figure 1 (A) Plain X-ray kidney, ureter and bladder showing multiple radio opaque density shadow on right side and single radio opaque shadow on left side at L1 level suggestive of renal stones. (B) Intravenous pyelogram showing bilateral renal stones with distorted dilated calyces. (C-H) CT images (axial and coronal cuts) showing extrarenal calyces with bilateral renal stones and right gross hydronephrosis suggestive of pelviureteric junction obstruction.

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To cite: Parmar KM, Tyagi S, Singh Skumar, *et al.* *BMJ Case Rep* 2019;**12**:e228684. doi:10.1136/bcr-2018-228684

I hydronephrosis with bilateral nephrolithiasis and abnormal pelvicalyceal system (figure 1B). CT urography showed bilateral extrarenal calyces and renal pelvis along with two renal veins on either side. (figure 1C-H). Metabolic stone work up including serum calcium, uric acid, parathyroid hormone and citrate were within normal limits.

Treatment

Patient was planned for staged procedure, dealing more symptomatic left side first. Percutaneous nephrolithotomy was performed and complete stone clearance achieved. Right side retrograde pyelography showed gross hydronephrosis, 'jet sign' at pelviureteric junction (PUJ) and delayed drainage. Postoperative course was eventful and patient was discharged on day 3. Stone analysis revealed mixture of calcium oxalate monohydrate and dihydrate stones. Patient was readmitted after 4 weeks for robot assisted laparoscopic pyeloplasty along with stone retrieval. Under general anaesthesia, patient was placed in right flank up position. Standard laparoscopic ports were placed and robot docking was done. Intraoperatively, colon was mobilised and gonadal vessels were traced up to the renal hilum (figure 2A,B). The dissection at hilum revealed early bifurcation of main renal vein, with a smaller inferior pole tributary crossing anterior to PUJ. Right gonadal vein was draining into inferior polar renal vein (figure 2C). The gonadal vessels were preserved. A hugely dilated baggy pelvis was seen connected to extrarenal calyces by finger-like projections. Multiple round stones were retrieved and standard Andersen-Hynes dismembered pyeloplasty was performed over 4.8 French double J stent with excision of fibrotic and obstructed PUJ segment, along with Hell storm stitch of anterior crossing vessel to the baggy pelvis (figure 2D-F).

OUTCOME AND FOLLOW-UP

Postoperative course was uneventful, and patient was discharged on day 4. Double J stent was removed after 6 weeks as a day care procedure. At 6-month follow-up, the patient was asymptomatic, with the diuretic renal scan showing adequate renal drainage and no recurrence of stones.

DISCUSSION

First reported by Eisendrath in 1925, extrarenal calyx is an extremely rare abnormality¹ and until today only about 20 cases have been documented in literature. The exact embryological basis remains to be elucidated; differential rate of development of metanephric ridge relative to rapidly maturing ureteric bud, resulting in prior development of calyceal system before merging with metanephric tissue is the most favoured hypothesis.² Extrarenal calyces may be associated with other congenital anomalies of urinary tract like renal ectopia, duplex kidney, horseshoe kidney and renal dysplasia.³ Apart from bilateral extrarenal calyces, anomalous drainage of right gonadal vein into right renal vein was present in this particular case. This anomalous drainage of right gonadal vein has been variously described in up to 1.5% of cases.^{4,5} Embryologically, supracardinal and subcardinal sinuses develop during early gestation. Persistence and regression of the right supracardinal sinus result in left gonadal vein draining into the ipsilateral renal vein and right gonadal vein and right renal vein draining into the inferior vena cava. Variations in relation to gonadal vein are more commonly seen on left side. Knowing these anatomic variations is essential for the urologist particularly operating on kidneys, suprarenal gland and retro peritoneum. The index case had PUJ obstruction secondary to an

anteriorly crossing accessory vessel and fibrotic PUJ area leading to multiple secondary renal stones. The incidence of pelviureteric junction obstruction (PUJO) with extrarenal calyces is extremely low and since the clinical presentation is classical to PUJO, high index of suspicion should be kept in case of distorted calyces on preoperative imaging studies. Knowing this, urologist would be cautious enough to safeguard and prevent injury to calyces and other anomalous vessel while operating on PUJO.⁶ The association of PUJO, extrarenal pelvis and calyces and anomalous gonadal vein drainage is rare. To best of our knowledge it has not been described earlier. Diagnosis of these anomalies requires detailed and adequately performed imaging studies. Plain radiography and ultrasonography offers limited information. CT urography provides good anatomic information like in our case, which shows long finger-like projections of extrarenal calyces draining into dilated pelvis. Management depends on renal function and patient's symptom. Asymptomatic incidentally detected extrarenal calyces without any other associated anomaly requires follow-up. PUJO with extrarenal calyces requires reconstruction in the form of pyeloplasty creating wide; tension-free mucosa to mucosa anastomosis and adequate drainage without injuring the extrarenal calyces. Wadhwa and Hemal reported transmesocolic laparoscopic pyeloplasty in a pelvic kidney with PUJO and extrarenal calyces.⁷ The index case had multiple secondary stones with PUJO. We opted for robot assisted pyeloplasty and could retrieve all the stones.

Learning points

- ▶ Pelviureteric junction obstruction is a common upper urinary tract congenital anomaly.
- ▶ Multiple upper urinary tract anomalies in a single patient is rare.
- ▶ Robot-assisted surgery assist in reconstruction.
- ▶ When operating on a renal moiety with abnormal calyceal appearance on preoperative imaging, it is important to bear the above various anomalies in mind to safeguard against inadvertent injury of the calyces and vascular supply.

Acknowledgements We sincerely thank the patient for being cooperative during the hospital stay.

Contributors KMP: planning, data collection. ST: data collection, design and conception. SS: interpretation, expert comments. GS: data analysis.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

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