Case Report

Upper moiety vascular ureteropelvic junction obstruction in an incomplete duplex kidney: A variant of the Fraley's syndrome?

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ABSTRACT

Although duplication of the collecting system is fairly common, ureteropelvic junction obstruction of the upper moiety in a patient with duplex system is rare, more so if the obstruction is extrinsic and vascular. Herein, we report a case of obstruction of the upper moiety infundibulum by a crossing vein, in a patient with partially duplex system who presented with flank pain and focal hydronephrosis of the superior calyx, a clinical presentation similar to that of Fraley's syndrome. The infundibulum was transected and transposed anterior to the crossing vessel.

INTRODUCTION

Duplication of the collecting system is a common congenital anomaly of the upper urinary tract with an incidence of 0.8%.[1] Incomplete duplication, where the confluence of the two separate ureters is located anywhere above the ureterovesical orifice, is slightly more prevalent than the complete duplication. Ureteropelvic junction obstruction (UPJO) is a common anomaly rarely observed in incompletely duplicated systems and more commonly affects the lower moiety. UPJO in the upper moiety is rare, more so when the obstruction is extrinsic and vascular. A similar case of obstruction of the upper moiety by a crossing vessel in a completely duplicated system, was described in 1978.[2] Here, we report a case of extrinsic obstruction of the upper renal moiety caused by a crossing vein, in a patient with bifid pelvis, which may be considered as a variant of the Fraley's syndrome, and its management.

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CASE REPORT

A 36-year-old male presented with the history of intermittent right flank pain for the past 3 years. Computed tomography urography revealed an under rotated right kidney with incomplete duplication of the collecting system, where the single major calyx of the upper moiety was draining into the pelvis of the lower moiety. The upper moiety was hydronephrotic and a renal vein could be identified crossing anterior to the junction of the upper moiety infundibulum with the pelvis of the lower moiety, resulting in obstruction [Figure 1a-c]. ^{99m}Technetium diethylenetriaminepentaacetic acid (^{99m}Tc-DTPA) scan revealed a glomerular filtration rate (GFR) of 33 mL/min with delayed clearance from the upper moiety [Figure 2a and b]. Retrograde pyelogram revealed incomplete duplication of the renal pelvis with hydronephrotic upper moiety and delayed drainage [Figure 1d]. Surgical intervention was planned in this patient in view of symptoms along with delayed clearance from the upper moiety. A robot-assisted laparoscopic approach using the Da Vinci Xi Robotic

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system was chosen under general anesthesia. After colon mobilization, the dilated upper pole moiety was identified through the Gerota's fascia, which was then opened and the dissection of the upper moiety pelvis was continued caudally to identify the crossing vein [Figure 3a]. Subsequently, the lower moiety pelvis and the upper 1 cm of the ureter were partially dissected free of the overlying Gerota's fascia and this approach avoided the need of ureter and lower pole mobilization and thus prevented any vascular injury to either of the structures [Figure 3b]. The renal pelvis was incompletely duplicated and a vein was crossing anterior to the infundibulum of the upper moiety, just proximal to its insertion into the lower moiety pelvis, causing obstruction and hydronephrosis of the upper moiety [Figure 3c and d]. The lower moiety pelvis and UPJ were normal. The upper moiety pelvis was transected via an oblique incision just proximal to the crossing vein and spatulated and an end-to-end anastomosis was performed after it was transposed anterior to the crossing vessel [Figure 3e and f]. A Double J (DJ) stent was placed across the anastomosis with its upper coil in the upper moiety. The procedure and the postoperative course was uneventful. The patient was discharged on day 3 and the DJ stent was removed 4 weeks later. At 3-month's follow-up, the patient was symptom free. Follow-up ultrasonography showed resolution of hydronephrosis and the DTPA scan showed a GFR of 32.5 mL/min with improved subrenal drainage of the affected moiety [Figure 2c, d and e].

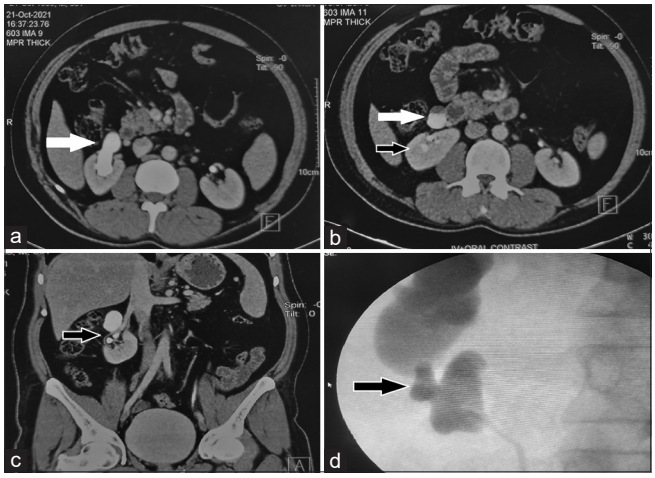

Figure 1: Axial (a and b) and coronal (c) contrast-enhanced computed tomography images showing malrotated right kidney with hydronephrosis of the upper moiety (a) with nondilated lower moiety calyceal system (b) and a crossing vein compressing the infundibulum of the upper moiety when it enters the lower moiety pelvis. (d) Retrograde pyelogram image showing dilation of the upper moiety and normal non dilated lower moiety pelvis. Exact level of obstruction is not appreciated as the kidney is malrotated

DISCUSSION

Normally, a single ureteric bud arises from the Wolffian duct and penetrates the metanephric mesenchyme and divides dichotomously to form the pelvicalyceal system. Incomplete duplication of the collecting system results when the ureteric bud divides prior to meeting the metanephros.[3] The abnormalities that affect a single renal collecting system such as UPJO have been shown to primarily affect the lower moiety of the duplex kidney; this is because the lower segment is the analog of a single renal system with usually about two-thirds of the renal parenchyma, at least 2 calyces and a true pelvis. This might explain the predilection of UPJO in the lower moiety.[4] The incidence of UPJO in a duplex kidney is 2%–7% and large series have reported a sigificantly lower incidence in the upper moiety (13%) as compared to the lower moiety (87%).[5] The upper segment

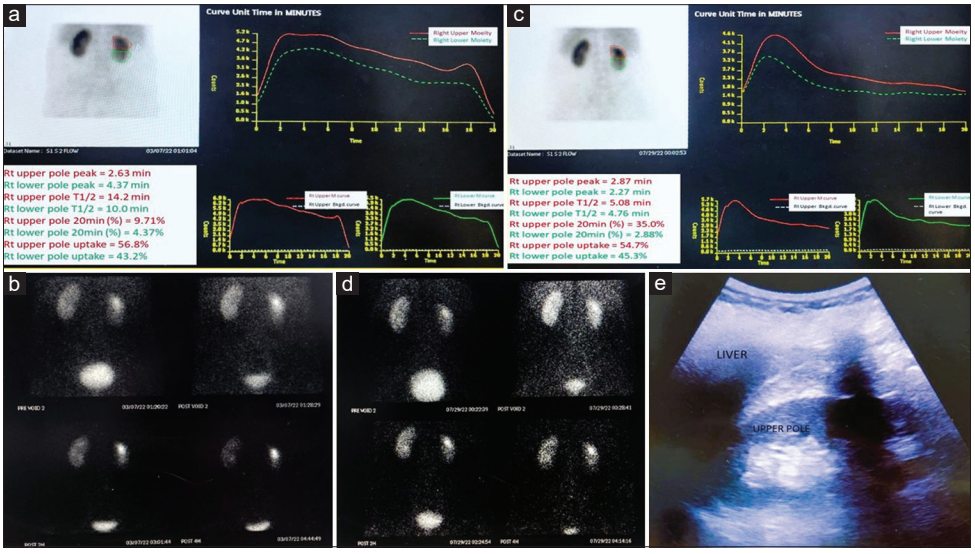

Figure 2: (a and b) Images of the preoperative DTPA scan: Both the upper and lower moiety curves show delayed clearance with uprising towards the end of the curve (a) and persistent tracer activity at the end of the 4 h (b). The curves are not very representative due to significant overlap in the region of interest chosen for DTPA, as the kidney was mal-rotated. (c and d) Images of the postoperative DTPA scan - Both the upper and lower moiety curves show a typical type I curve without any plateau or uprising at the end (c) and the significantly reduced tracer activity at the end of 4 h (d). Note the significant reduction in T1/2 in the postoperative curve. (e) - post operative ultrasonography image showing resolution of upper moiety hydronephrosis. DTPA: Diethylenetriaminepentaacetic acid

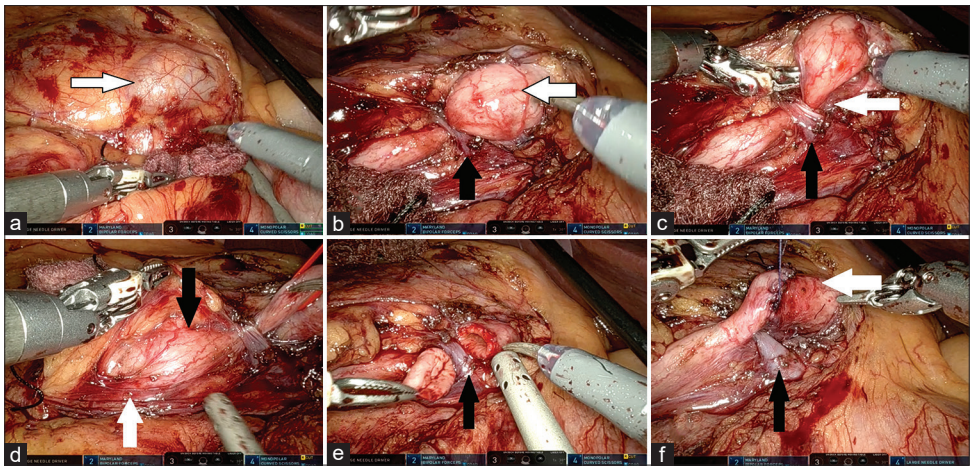

Figure 3: (a) Intraoperative picture-the colon has been mobilized and the upper moiety pelvis is visible through the Gerota's fascia (white arrow). (b) The dilated upper moiety pelvis (white arrow), the crossing vein (black arrow) and the nondilated lower moiety pelvis are seen. (c) The dilated upper moiety pelvis is lifted to show the normal diameter infundibulum (white arrow) under the crossing vein (black arrow). (d) The upper moiety pelvis, crossing vein, nondilated lower moiety pelvis (black arrow) and the single ureter (white arrow) are seen. (e) The upper moiety pelvis is transected in an oblique fashion and the crossing vein is transposed posteriorly (black arrow). (f) Post-anastomotic image (white arrow) showing the posterior transposition of the crossing vein (black arrow)

usually consists of one major calyx drained by a single infundibulum, and no true UPJ exists. The cases of upper pole UPJO are probably more analogous to infundibular stenosis or a calyceal diverticulum,[6] and thus, some authors prefer the terminology "upper-pole calyceal obstruction" or just "upper-pole obstruction." Irrespective of the arguments about the terminology, obstruction at the upper moiety of a duplex renal system does occur.[6] Furthermore, the majority of the UPJO in duplex systems are caused by intrinsic obstruction and crossing vessels are rarely reported to cause of UPJO in duplex kidneys. Thus, the presentation of UPJO in the upper moiety of a partially duplex system, that too because of an extrinsic vascular obstruction, due to a crossing vein, is rare and the authors are unaware of such a report published in the English literature. The differential diagnosis of the radiographic appearance of an obstructed moiety of a duplex kidney includes segmental cystic changes and extrarenal retroperitoneal lesions.[6]

Fraley's syndrome, first described in 1966, is extrinsic vascular compression of a calyceal infundibulum caused by a branch of the renal artery, most commonly affecting the superior calyx in a kidney with a single collecting system.[7] In 1978, Koyanagi described the first and the only reported case of Fraley's syndrome in a completely duplex kidney resulting from the compression of the upper moiety infundibulum by a branch of renal artery against the posterior renal vein.[2] The patient underwent ureterolysis and repositioning of the transected ureter anterior to the crossing renal vessel followed by an end to side uretero-ureterostomy of the upper moiety ureter to the lower moiety.

Our patient had partially duplex system, with obstruction of the upper moiety infundibulum by a crossing vessel; however, the obstruction was extrarenal instead of intrarenal in the classical Fraley's and the vessel was a vein instead of an artery, but still can be considered as a variant of the Fraley's syndrome. The pelvis of the upper moiety was transected just above its insertion into the common pelvis and an end-to-end anastomosis was made after transposing it anterior to the obstructing vein, similar to what is descried for Fraley's syndrome. The other surgical options in patients with upper moiety UPJO are pyeloureterostomy, pyeloplasty, or heminephrectomy, depending on the extent of duplication and the moiety's function.[6]

CONCLUSION

UPJO affecting the upper moiety of a duplex kidney is a rare entity, and such an obstruction when caused by an extrinsic vessel is even rarer. When managing UPJO in duplex systems, the possibility of an extrinsic anatomical cause of obstruction should be considered and looked for.

Declaration of patient consent

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

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