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Congenital right sided ureteropelvic junction obstruction in right crossed fused ectopia with extrarenal calyces masquerading as massive retroperitoneal urinoma in a case of blunt trauma abdomen: A diagnostic enigma and novel approach of management

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KEYWORDS

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Abstract

An 18 years old young male presented with history of blunt trauma to abdomen with mild hematuria. The contrast enhanced computerized tomography of abdomen revealed crossed fused right renal ectopia with distal large fluid filled sac confusing with urinoma. A midline laparotomy revealed it to be case of right hydronephrotic ectopic fused kidney with extrarenal calyces with large midline pelvis in retroperitoneum. Tailoring of renal pelvis with preservation of posterior pelvic wall plate and ureteral reconstruction was done which was anastomosed to native right ureter. The patient is doing well in one year of follow-up.

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Case summary

An 18-year-old young male presented with history of blunt trauma to abdomen followed by mild hematuria for one week. Abdomen was mildly tender. His routine microscopic urine examination showed plenty of red cells. The ultrasound of abdomen showed crossed fused right ectopic kidney lying low in left lumbar region with a fluid filled large round sac distal to kidney. The renal function tests were normal. The CT scan abdomen showed crossed fused right ectopic kidney with large sac containing urine just distal to the fused kidneys (Fig. 1A, and drawing-1). A retrograde ureteropyelogram of right side was done showed contrast accumulation in the sac starting from dome of bladder and the pelvicaliceal system could not be clearly delineated. The left pelvicaliceal system was normal.

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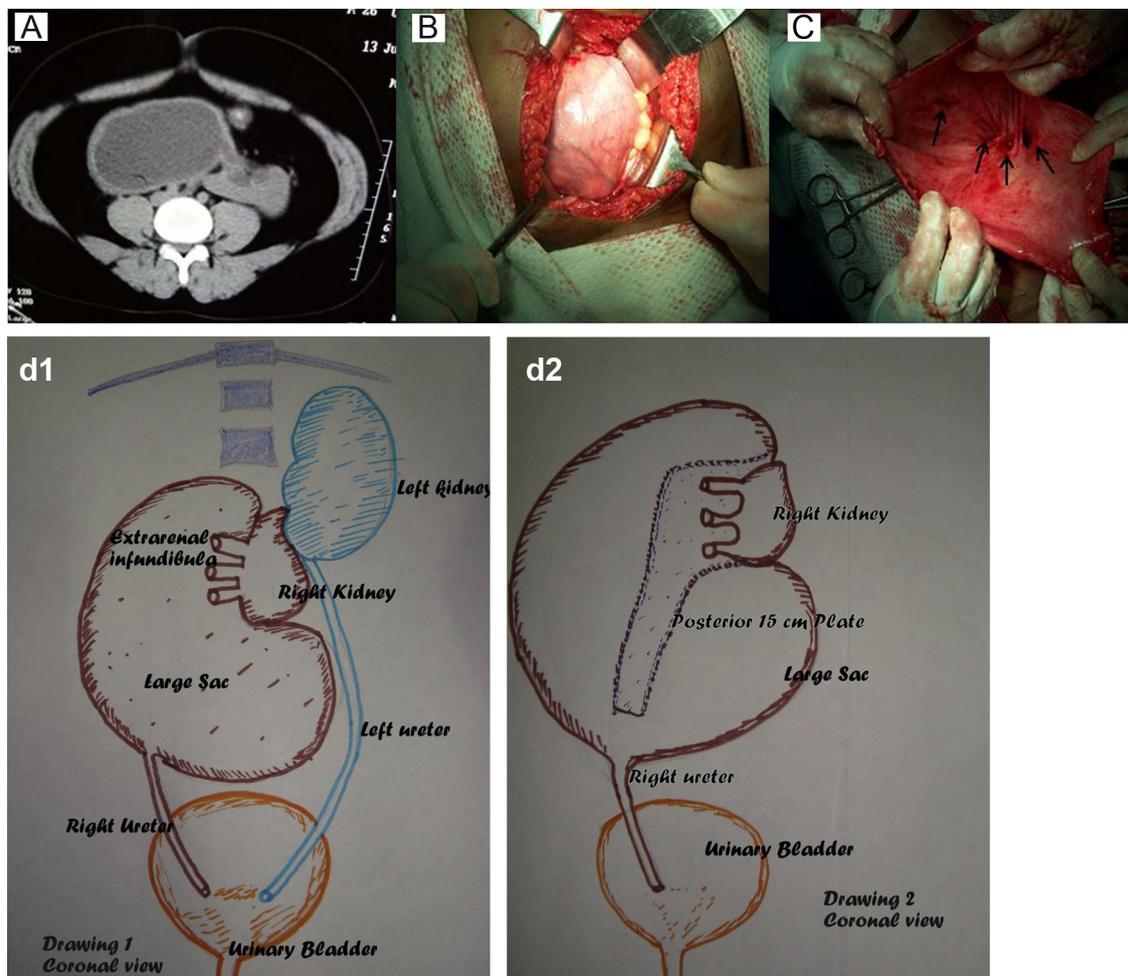


Figure 1 (A and drawing-1) CT scan abdomen showing large sac connected to pelvi-caliceal system of ectopic kidney. (B and drawing-2) The large fluid filled sac in retroperitoneum. (C) Opened up pelvic cavity showing lumens of calyces.

With the clinical and radiological diagnosis of urinoma secondary to possible ureteric injury, the patient was subjected to infraumbilical midline laparotomy. Intraoperative finding was a large fluid filled sac lying in centre of retroperitoneum (Fig. 1B and drawing-2). The ureter was identified and its junction to the sac was adherent to surroundings were dissected. It showed stenotic and aperistaltic UPJ lying low just posterior to the dome of bladder on right side. The opened sac showed openings of calyces classical of extrarenal calices (Fig. 1C). There was large gap between the UPJ and the ectopic kidney. A 15 cm posterior wall of sac was preserved with its vascular sheath and whole of the sac was trimmed. This posterior plate was reconstructed like upper ureter over 6F double J stent by 4-0 polygalactin interrupted sutures (Fig. 2A and drawing-3). This reconstructed ureter was anastomosed end to end to the spatulated native right ureter (Fig. 2B and drawing-4). The postoperative course was uneventful. The biopsy of the excised UPJ segment showed defective circular muscle fibres with excessive collagen deposition, consistent with UPJ obstruction. The stent was removed after 8 weeks. A RGP done following stent removal showed patent ureter with no contrast extravasation (Fig. 2C). Intravenous urogram at 6 months showed no hydronephrosis. A DTPA renogram showed non-obstructed drainage. The patient is doing well in 12 months of follow-up.

Discussion

Extrarenal calyces are uncommon congenital anomaly in which the major calyces as well as renal pelvis lie outside the renal parenchyma. They usually do not produce symptoms although failure of normal drainage can lead to stasis, infection and calculi formation. This entity can co-exist with ectopic and horseshoe kidneys. The ureteral atresia and renal dysplasia are the other associations with this anomaly [1]. The association of primary ureteropelvic junction obstruction with ectopic kidney has already been described but the association of extrarenal calyces with concomitant UPJ obstruction has not been reported earlier. The mechanism of hematuria in blunt trauma in UPJ obstruction is due to the rupture of suburothelial vessels and in most of cases it is self limiting and responds by conservative treatment [2]. The extrarenal calyces and infundibulum in association with hydronephrosis was described first by Eisendrath D.N. in 1925 and 20 cases so far have been reported in English literature [3,4]. The exact aetiology of extrarenal calyces is not very clearly understood. It has been hypothesized that the anomaly could be due to a disparity resulting from slow development of the metanephric tissue or to a relatively rapid development of ureteric bud. If the ureteric bud has a rapid or a precocious development, the calyceal system could well develop prior to its coalescence

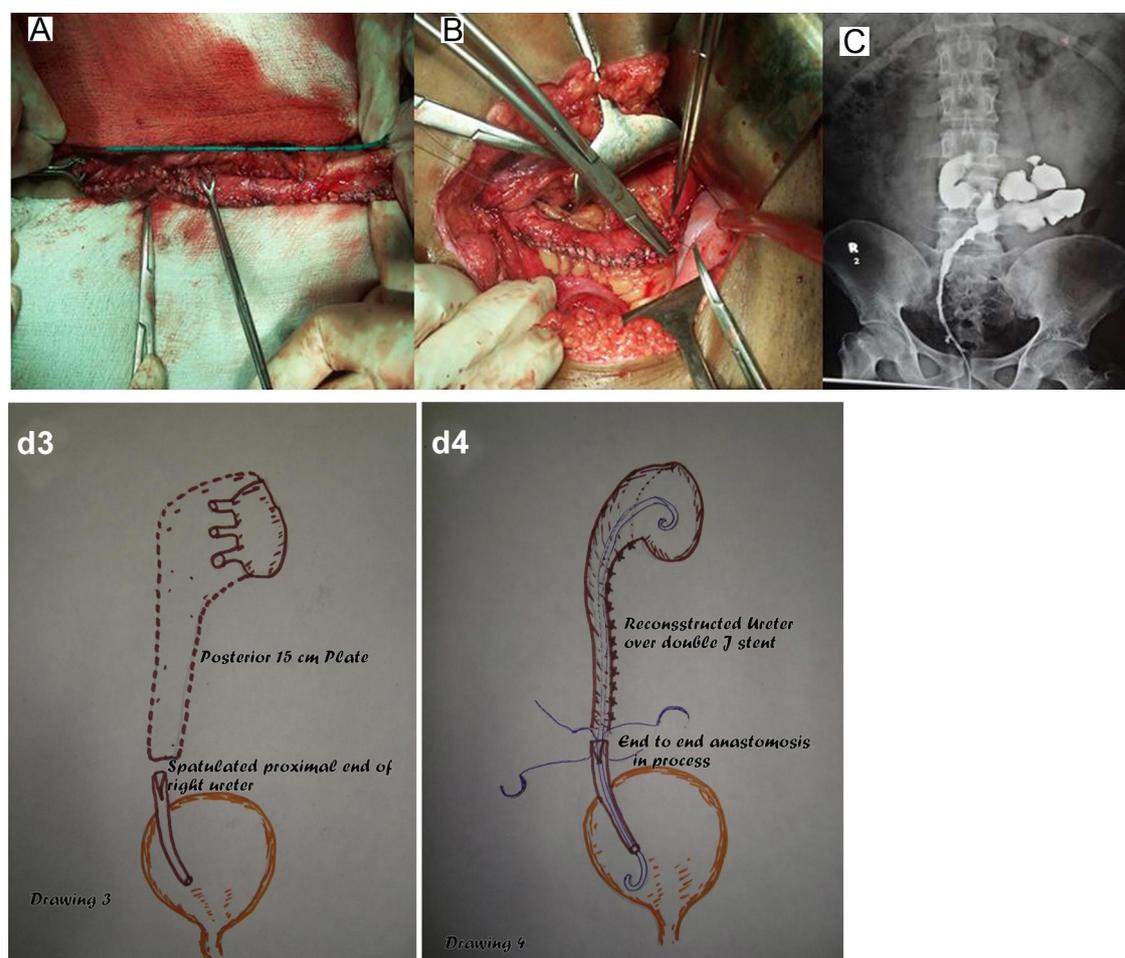


Figure 2 (A and drawing-3) Reconstruction of upper ureter from posterior pelvic wall plate. (B and drawing-4) End to end anastomosis of proximal reconstructed ureter to distal native right ureter. (C) Retrograde uretero-pyelogram showing reconstructed ureter.

with the nephrogenic mass. Conversely, lag in the growth of nephrogenic mass could delay its attachment to the collecting system permitting extrarenal development of the first or second order of the collecting system [5,6]. The extrarenal calyces could remain undetected for long time or diagnosed because of the complications arising from it. The complications could be the infection, stones or stasis due to the anomaly [7,8]. The presence of the associated anomalies such as ectopia, bifid system, renal dysplasia and presence of horseshoe kidneys influence the outcome and prognosis [9]. In present case, the UPJ obstruction was present with the anomaly and trauma caused hematuria which compelled the patient to attend hospital and the anomaly was diagnosed intraoperatively. The preoperative CT scan with clinical background, a possibility of ureteric injury could not be ruled out and a most probable diagnosis of urinoma was made. The ureteric reconstruction over 6-F ureteric stent was done from posterior plate of the hydronephrotic pelvic sac and it was anastomosed to the native ureter lying low in the pelvis on contralateral side. The present case is unique in the sense that the extrarenal calyces were associated with UPJ obstruction which has not been reported earlier. The presentation of patient was with hematuria due to blunt trauma of abdomen and large hydronephrotic kidney with pelvis confused the presentation to be the ureteric injury leading to urinoma formation. The CT scan was consistent with the possibility of ureteric injury and urinoma formation. The

bridging of large gap between the extrarenal calyces was done using the pelvic plate which was reconstructed like upper ureter. The follow-up DTPA scan showed non-obstructive drainage.

Conclusion

Be aware of rare anomalies of collecting system while evaluating a case of retroperitoneal urinoma after trauma. On exploration what one was thinking of urine extravasation, could be a rare collecting system anomaly. Dealing with such cases demands prompt recognition and appropriate surgical correction.

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