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"Duplex sans duplex: a cryptic cause": a case report

S. Rajkiran Raju[1], Jobin Pathrose[1], Dyan D'Souza[1] and Attibele Mahadevaiah Shubha[1*]

Abstract

Background Duplex kidneys represent an embryologic maldevelopment at time of renogenesis resulting in a spectrum of bifurcation anomalies of the reno-ureteric system. Though most are antenatally detected, recurrent urinary tract infections (UTIs), abdominal mass due to obstruction and incontinence are other common manifestations. Upper moiety ureter is usually obstructed and the lower moiety is refluxing. Management is guided by the percentage function of each of the moieties. A non-functioning system warrants a heminephrectomy. We report a toddler with right flank mass and a provisional diagnosis of right duplex system following investigations but met with a cryptic cause at surgery thereby altering the management.

Case presentation A 2 ½ years girl with progressively increasing right abdominal mass for 6 weeks was found to have 12 × 10 cm right non-tender flank mass. Ultrasonography, contrast tomography and nuclear scans showed a right duplex system with obstructed, poorly functioning lower moiety. A lower moiety heminephrectomy was planned but at surgery, a densely adherent cystic structure displacing the right kidney superiorly was noted. On decompressing, the ureter was found to enter the cyst with discontinuation for a length of 6cms before being traced distally to its entry into the bladder. Retrograde pyelogram confirmed mid-ureteral transection and cystic urinoma. The cyst was excised and the ureter reconstructed with an appendicular interposition graft. Child recovered uneventfully and at 8 months follow up is well with good drainage across the conduit.

Conclusion The case highlights a rare presentation of mid-ureteral transection with urinoma masquerading as a duplex system and its satisfactory management.

Keywords Duplex kidney, Ureteral transection, Urinoma, Appendix interposition

1 Introduction

Duplex kidneys comprise a spectrum of bifurcation anomalies of the reno-ureteric system. They commonly present with recurrent urinary tract infections (UTIs), abdominal mass due to obstruction or incontinence. Post-traumatic isolated ureteral injuries are extremely rare in children, due to anatomic concealment in the retroperitoneum. Ureteral transections following blunt abdomen trauma result in urinomas, often manifesting acutely with fever and toxicity due to early infection or with an abdominal mass. Management of ureteral injuries depend on the level, extent and completeness of disruption. Complete ureteral transections mandate surgical restoration of continuity.

We herewith report a rare presentation of isolated right ureteral transection with large urinoma in a two and half-year-old girl following unnoticed trivial trauma masquerading as a duplex right kidney with lower moiety obstruction and highlight the challenges encountered in management.

*Correspondence:
Attibele Mahadevaiah Shubha
dramshubha@yahoo.co.in
[1] Department of Pediatric Surgery, St Johns National Academy of Health Sciences, Bangalore, Karnataka, India

2 Case presentation

A two and-a-half-year-old girl was brought to hospital, with the mother noting a progressively increasing mass in the right abdomen for 6 weeks. She was otherwise well, but for two spells of low-grade fever which resolved with antipyretics. She weighed 10.2 kg, was afebrile and normotensive (Blood pressure 50th centile). A firm, non-tender 12×10 cm mass in right lumbar region extending to the right hypochondrium, abutting the anterior abdomen wall demonstrating right flank fullness with dullness was noted on abdominal examination. Genitalia, spine and rest of the systemic examination were unremarkable.

She was evaluated elsewhere with an abdominopelvic ultrasonography (USG) showing a right duplex kidney with mild hydronephrosis of the upper moiety and a large hydronephrotic lower moiety without ureteric dilatation. The contralateral left kidney was normal. A contrast-enhanced computed tomography (CECT) (Figs. 1, 2, 3A) revealed a large (89×83 mm) homogenous mass with peripherally enhancing rim without any contrast excretion (white arrow) at the lower part of mildly hydronephrotic right kidney (black arrow). The mass extended cephalad into subhepatic region, displacing the inferior vena cava (IVC) medially. The ureter was not delineated on right side, the contralateral left kidney and ureter were normal suggesting possible duplex right kidney with poorly functioning hydronephrotic lower moiety. The other possible differentials for pediatric flank mass with the above CT findings include cystic disease of kidney, retroperitoneal lymphatic cyst and tumors like Cystic nephromas.

Micturating cystourethrogram (MCUG) showed normal bladder contour and absent post-void residue (PVR) without vesicoureteral reflux (VUR) (Fig. 3B). Ethinyl di- Cysteine (EC) diuretic renogram revealed a small right upper moiety with functional stasis showing progressive clearance and a large photopenic area below, with poor tracer uptake and obstructed drainage. The left kidney showed good uptake, normal intrarenal transit with unobstructed tracer excretion. The relative function of left:right was 78:22 with the right lower moiety contributing to only 12% of right kidney function (2.6% of overall renal function) (Fig. 4). Her complete haemogram was normal, with Serum creatinine of 0.4 mg% and eGFR- 92 ml/min/1.72m^2. Urine routine/microscopy and culture were sterile.

Based on the clinical findings and imaging, a provisional diagnosis of duplex right kidney with grossly hydronephrotic, obstructed, poorly functioning lower

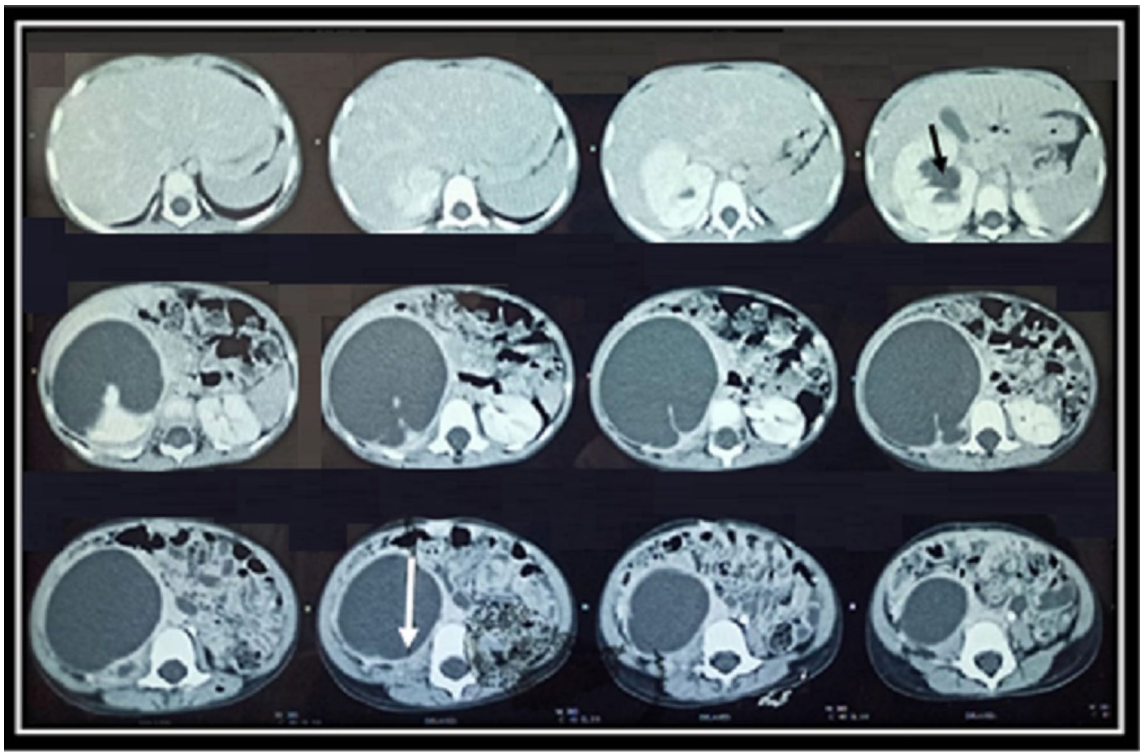

Fig. 1 Axial tomogram of CECT Abdomen showing right mild hydronephrosis (upper moiety) (black arrow), peripherally enhancing cystic mass at lower pole (white arrow) suggestive of duplex right kidney

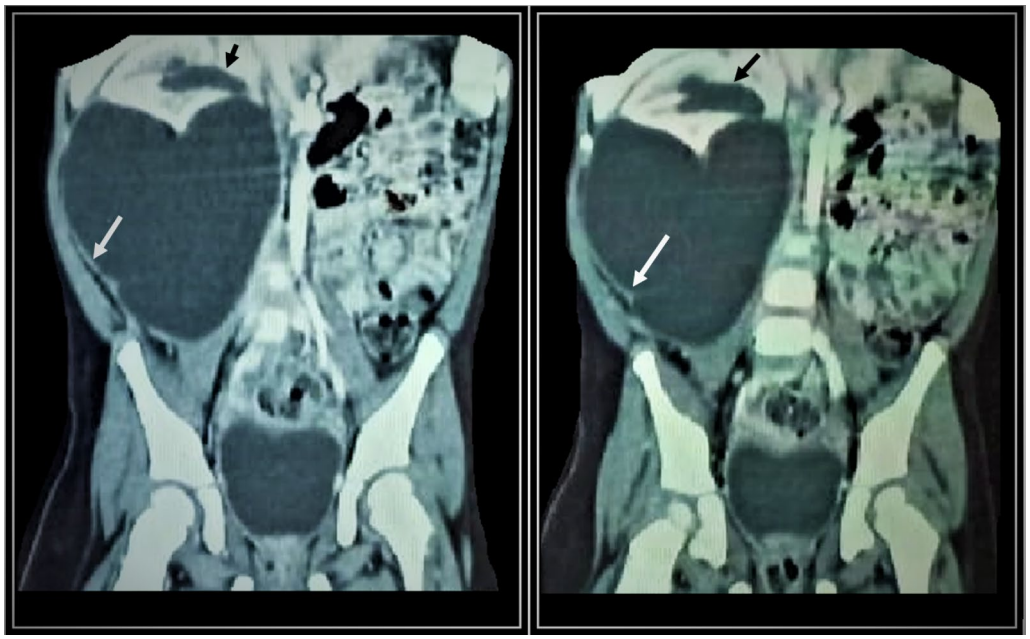

Fig. 2 Coronal reconstruction images of CECT Abdomen showing features suggestive of right duplex kidney with mild upper moiety hydroureteronephrosis (black arrow) and thin walled (white arrow) hydronephrotic right lower moiety without contrast excretion

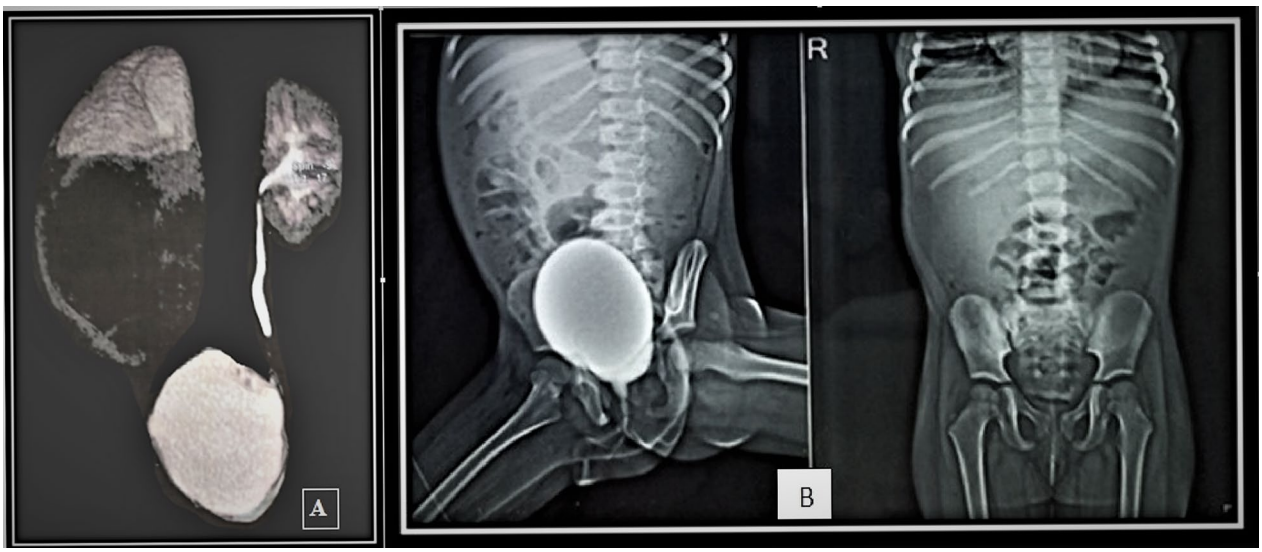

Fig. 3 Reconstructed CECT image showing right lower moiety hydronephrosis with thin rim of parenchyma (**A**). MCUG (**B**) normal bladder contour without VUR and absent PVR

moiety was made and an open right lower moiety heminephrectomy was planned.

At laparotomy, the upper pole of right kidney was displaced cephalad onto lower surface of liver. The lower moiety which measured 14×10 cm (Fig. 5A) was found to be densely adherent posteriorly to the retroperitoneum, 11th and 12th ribs and medially to the mesentery of colon, duodenum and the IVC. Upon decompression, 750 ml of clear urine was evacuated. A single ureter of right side was noted to insert into the lower pole of this collection (Fig. 5A). The wall of the cystic collection was deroofed and excised all around except for a sliver of tissue along the IVC. Post excision, urine was noted to drain from a small opening on lower surface of the right kidney (Fig. 5, red arrow). An on-table Retrograde pyelography (RGP) showed the mildly hydronephrotic pelvicalyceal

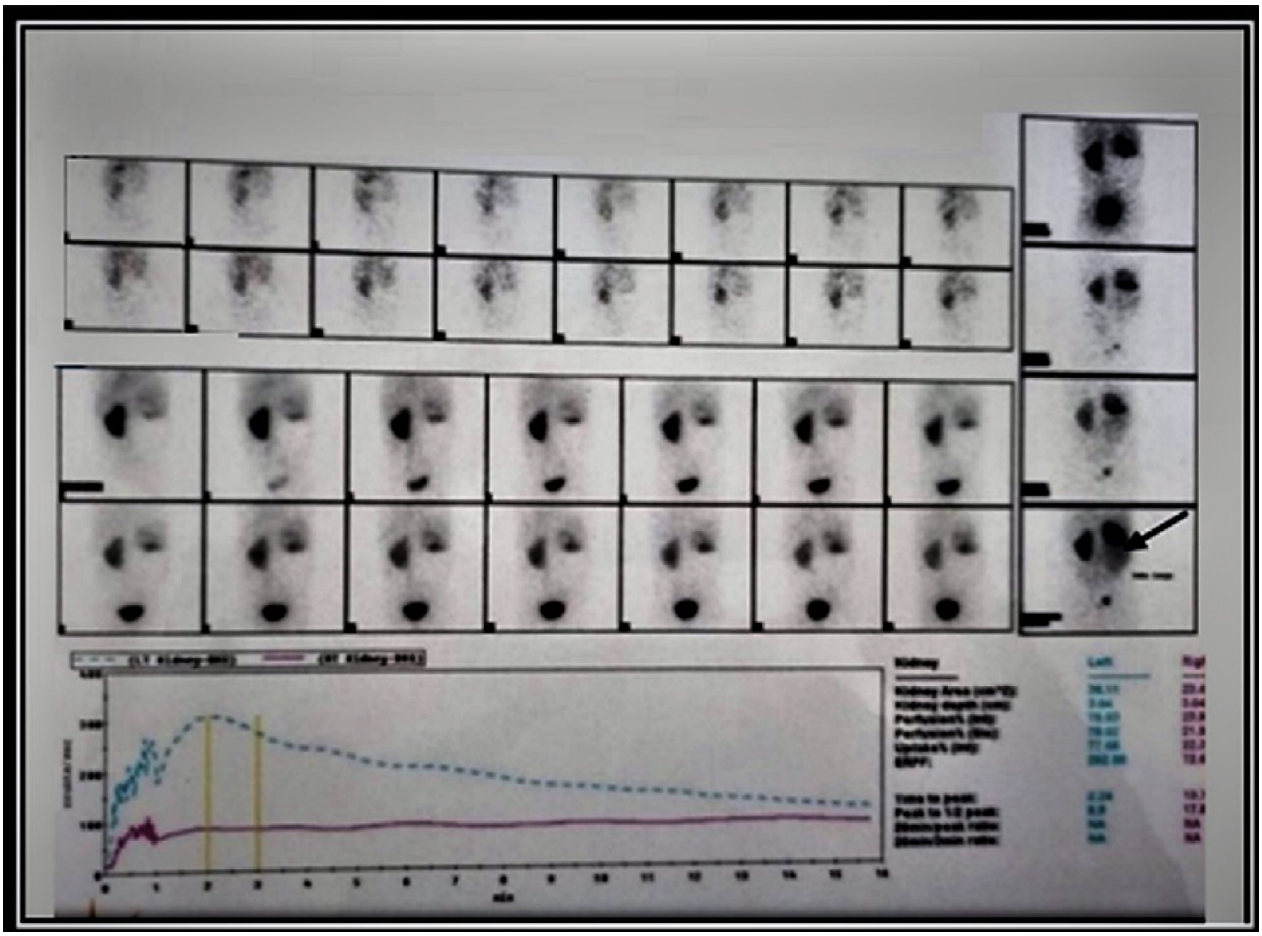

Fig. 4 EC renogram—normal left kidney and ureter with unobstructed drainage. Right duplex with obstructed right lower moiety in delayed film (black arrow)

system (PCS) with short upper ureter (Fig. 5B). The ureter distally from the collection to bladder was patent (confirmed by passage of 6 Fr Infant feeding tube and saline flushing). The gap between the upper and lower ureter after excising the cyst wall was around 6cms.

A pedicled appendicular interposition graft (Fig. 6A, B) was harvested and end to end anastomosis with upper and lower ureter was done over 3.5 Fr-18 cm Double J (DJ) stent. The position of the DJ was confirmed by an on-table fluoroscopy (Fig. 6C).

Postoperatively, the child recovered uneventfully. The histopathology of the excised wall showed no identifiable renal parenchyma, but only dense interstitial tissue with fibrosis, confirming, *the final diagnosis* of right lower polar perinephric urinomas following complete mid-ureteral transection.

In retrospect, on questioning, the mother could barely recollect the trivial fall of the child from edge of the bed 6 weeks back. Though not more details than this were recounted.

She was discharged on uro-prophylaxis for 6 weeks followed by cystoscopic DJ stent removal. An RGP showed a patent appendicular interposition graft without leak (Fig. 7A). A diuretic EC renogram after three months post stent removal showed prompt unobstructed drainage, improvement in right renal function (Relative function Right: Left: 38:62) with improved parenchymal perfusion, uptake and intra-renal transit times (Fig. 7B). The child on follow-up 8 months post-surgery is asymptomatic, voiding well and is off all medications. An interval renal scan during this visit showed complete resolution of right hydronephrosis.

3 Discussion

Reno-ureteric duplication is defined as a single renal unit with two pelvicalyceal systems and/or ureter(s); is one of the common anomalies of the genito-urinary tract with an estimated incidence of 1% of live births and a definite female predilection [1]. Although diagnosed antenatally, delayed or missed cases are not uncommon. Common

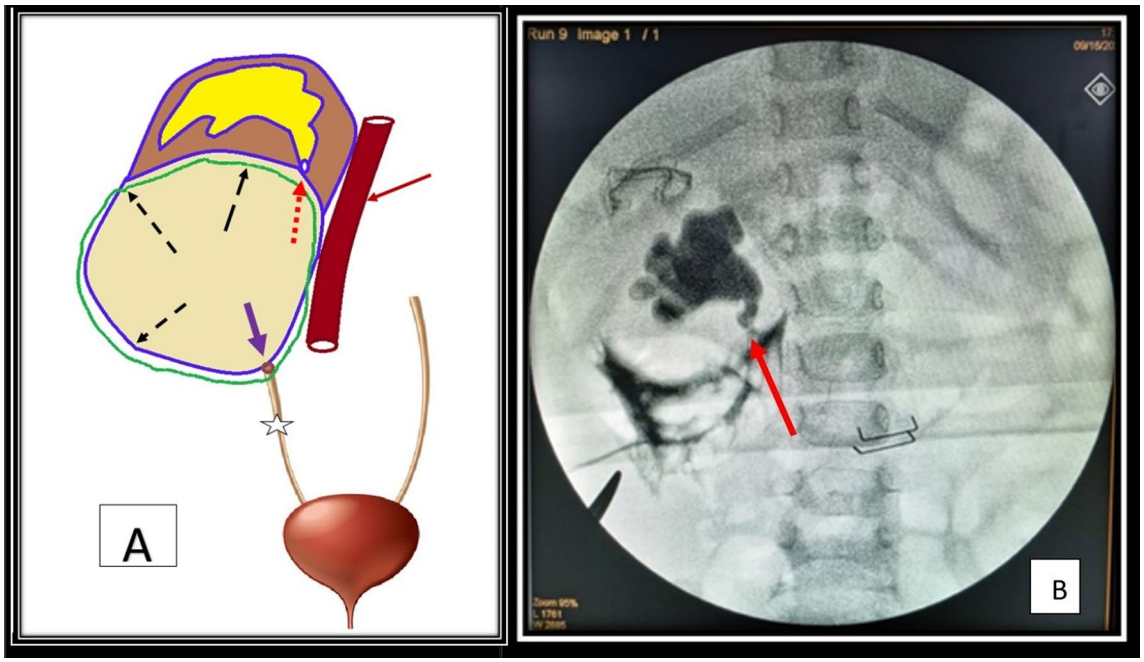

Fig. 5 **A** Intra-operative findings demonstrated by line diagram—Brown line shading represents upper moiety parenchyma, dotted red arrow—upper moiety ureteral opening, thinned out lower moiety (dotted green lines) with dotted black arrows indicating hydronephrosis. The point of communication of ureter with the collection (purple arrow) and inferior vena cava (dark red arrow). **B** Retrograde pyelography—mildly hydronephrotic upper moiety ureter opening (red arrow)

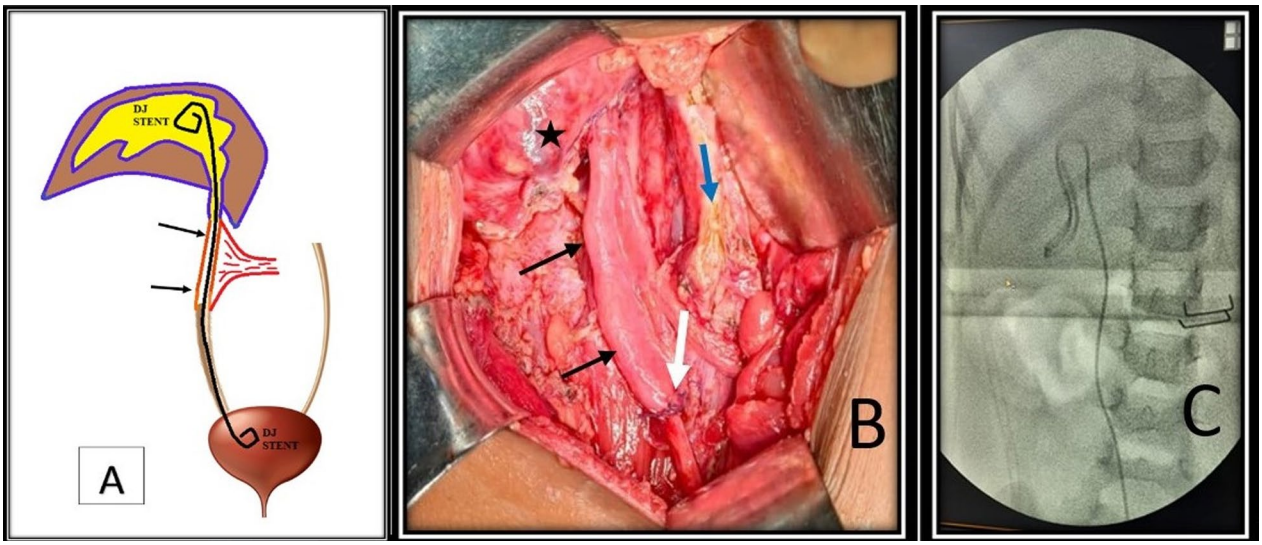

Fig. 6 **A** Line diagram **B**—corresponding intra-operative image—appendicular interposition graft (black arrow), inferior pole of right kidney (black star), Inferior vena cava (blue arrow) and anastomosis with lower ureter (white arrow). **C** On-table fluoroscopy image post anastomosis demonstrating the DJ Stent in situ

manifestations include recurrent urinary tract infections (UTIs), urinary incontinence, flank pain and flank mass. Duplex kidneys commonly become symptomatic as a result of upper moiety obstruction, VUR, incontinence due to ectopic ureters (females) and ureterocele [2]. Conventionally, imaging is by USG followed by MCUG to rule out VUR. Intravenous pyelography (IVP) has largely been replaced by radionuclide isotope diuretic renogram. Anatomic delineation especially in ectopic ureters is best with Magnetic resonance urogram (MRU) [3].

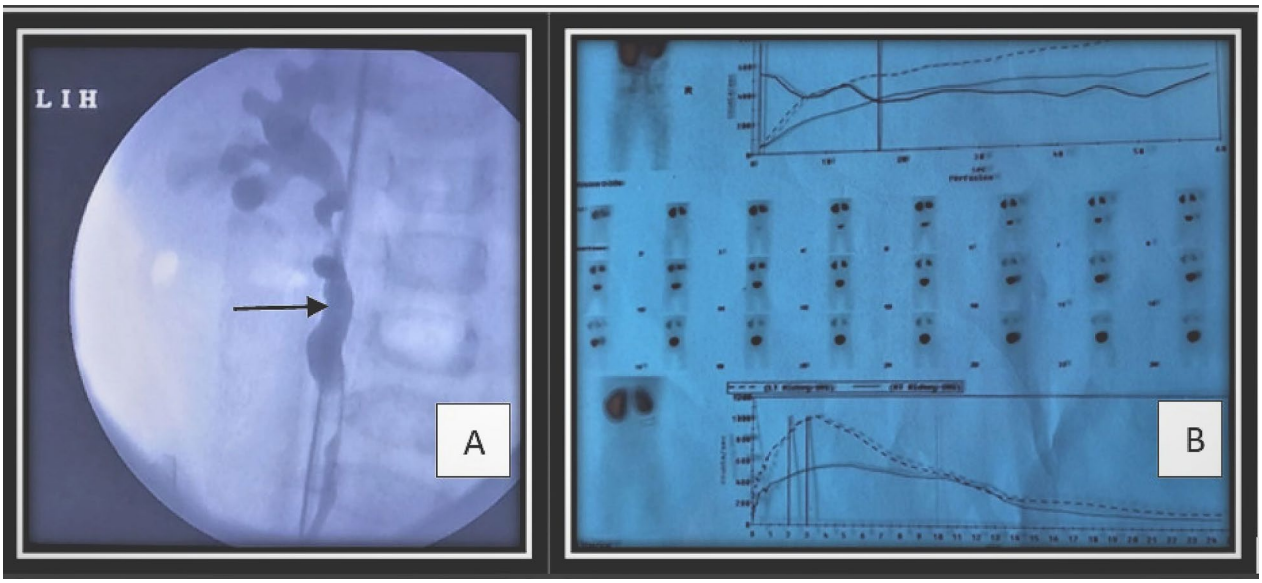

Fig. 7 **A** RGP post double J stent removal—site of anastomosis with free flow of dye (black arrow), **B** diuretic renogram post-surgery—improved perfusion and unobstructed drainage in the right kidney

Surprisingly, the duplex ureters fairly follow the embryologic lineage as described by the Weigert Meyer Law.

Surgical management of reno-ureteric duplications depend on laterality, relative function of each moiety, evidence of obstruction and presence of ectopic ureters/ureteroceles. Asymptomatic bifid renal pelves without VUR may not need intervention. An obstructed functional upper moiety due to pelvi-ureteric junction obstruction can be salvaged with upper moiety dismembered pyeloplasty while a non-functional system may be resorted to an upper moiety heminephrectomy. Ureteroceles without ectopic ureters may be treated with cystoscopic deroofing/puncture but ectopic ureters require diversion or ureteric reimplantation [4]. In our child, the unexpected intra-operative findings of ureteral transection with perinephric walled off urinomas ruled out reno-ureteric duplication contrary to the provisional pre-operative diagnosis.

Ureteral trauma is relatively infrequent in children; accounting for less than 1% of all urological traumas [5]. Isolated ureteral injuries resulting from blunt abdominal trauma are exceedingly rare due to anatomical concealment in the retroperitoneum by the pelvic girdle, iliopsoas muscle and vertebrae. Iatrogenic Ureteral injuries are commoner in adults and the incidence of penetrating abdominopelvic injuries from gunshot is on the rise in western population [6]. Pediatric renal injuries are common, accounting for 10–20% of blunt abdominal injuries in children; often co-existing with anatomic pathologies like pelvi-ureteric junction obstruction (PUJO), horseshoe or ectopic kidneys. Relative lower position of the pediatric kidneys with immature ribcage predisposes to such injuries [7]. Trauma may bring to fore the underlying unrecognized anatomical anomaly including tumors and attribute to symptoms including flank pain and hematuria [8]. The bladder and urethral injuries are more obvious and are commonly associated with pelvic fractures.

The ureteral injuries are classified by the American Association for the Surgery of Trauma [9] (AAST) into 5 grades with Grade I injury representing mild hematoma or contusion without devascularization, Grade II and III involving ureteral lacerations less than or more than 50%, respectively. Grade IV and V injuries involve lacerations with devascularization (< 2 cm or > 2 cm, respectively). The ureter being a retroperitoneal structure securely nestled by the bony pelvis, psoas muscle, the ligament of Trietz with inferior mesenteric artery and sigmoidal vessels on the left side. On the right, the ureter lies posterior to duodenum, IVC and ileocolic/right colic vessels. This implies that ureteral injury may be associated with severe life-threatening intra-abdominal injuries including hepatic, mesenteric and bowel tears [10]. These obvious findings may mask actual ureteral injuries. PUJ injuries present acutely with pain abdomen and progressively increasing mass with signs of toxicity from secondarily infected urinoma. In contrast, ureteral transections without other significant intra-abdominal injuries may be silent with gradual often self-limited retroperitoneal collection [11] as was in our case.

While ultrasound gives good idea about intra-abdominal solid organ involvement especially as a handy bedside screening modality, its use in imaging retroperitoneal structures in an emergent unprepared patient may be limited due to bowel gas. No single modality has been noted to be superior in diagnosing isolated ureteral injuries; a combination of CECT or RGP or a delayed phase IVP is recommended for maximizing the diagnostic accuracy of ureteral injuries [12].

Management of post-traumatic urinomas is challenging. Small urinomas with AAST Grade I and II may be managed conservatively. Ureteral disruption involving > 50% of circumference mandate surgical exploration, repair and restoration of anatomic continuity. These urinomas are notorious for dense retroperitoneal fibrotic adhesions and scarring [13]. Surgical management entails meticulous dissection across difficult dense adhesions and identification of transected ends of the ureter. Mid-ureteral short segment losses (< 2 cm) may be managed by mobilization with primary repair over DJ stent. Lower ureteral injuries may be managed by ureteric reimplantation with a bladder hitch procedure like Boari flap. Long segment ureteral losses require replacement with interposition grafts like vascularized appendix or ileum (based on Monti's principle) [14].

This case report highlights the rarity of isolated high grade mid-ureteral transection in a child with non-forthcoming history of trivial trauma manifesting as a large right flank mass and imaging studies suggesting ***right duplex system with obstructed lower moiety***. Intra-operatively, the presence of dense retroperitoneal fibrosis, a single ureter with mid ureteral loss and histopathology of the resected cyst wall confirmed the unexpected diagnosis of missed ureteral injury and urinoma. Appendix is a robust easily harvestable conduit to bridge long gap right ureteral defects with minimal complication rates and good long-term outcomes. A literature search of EMBASE, SCOPUS, PUBMED and Google Scholar did not reveal any such similar case of an isolated ureteral transection masquerading as a duplex kidney; hence, we present it as a protean case highlighting the use of appendicular interposition graft.

4 Conclusion

Renal masses in children are common and presentation is variable; anatomical deviations derive from aberrations in embryologic lineage. Accurate anatomical imaging is useful to delineate such aberrant anatomy. Perinephric urinomas may result following trauma, and presentation is classical with pain, sepsis and acute toxic signs but is not universal. In children, it may present as a flank mass with trivial symptoms. Retrograde pyelography is an excellent aid in delineating difficult uretero-pelvic anatomy. Vermiform appendix is a robust conduit for long segment ureteral reconstruction.

Abbreviations

UTIs	Urinary tract infections
USG	Ultrasonography
CECT	Contrast enhanced computerized tomography
IVC	Inferior vena cava
MCUG	Micturating cystourethrogram
PVR	Post void residue
VUR	Vesicoureteral reflux
EC	Ethylene di cysteine
RGP	Retrograde pyelogram
PCS	Pelvicalyceal system
DJ	Double J
IVP	Intravenous pyelogram
MRU	Magnetic resonance urogram
PUJO	Pelvi-ureteric junction obstruction
AAST	American Association for the surgery of trauma

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Author contributions

Dr RK/JP/DD—collated the history, findings, imaging data and investigations of the patient. Dr RK/ DD—review of literature. Dr RK—preparing the initial draft and preparing the representative line diagrams. Dr AMS—analyzing the draft and final editing. Dr AMS/ RK/DD/JP—involved in the surgery and management of the child. All authors have read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on request.

Declarations

Ethics approval and consent to participate

The study is approved by St Johns Medical college Ethics committee (IEC No – 11/23). Parents have consented for participation in the study.

Consent for publication

Parents have consented for publication of the report.

Competing interests

The authors declare that they have no competing interests.

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