

Ureteral Triplication With Ureteropelvic Junction Obstruction Type Of Article: Case Report

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Citation

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Abstract

Ureteral triplication is one of the rarest malformations of the upper urinary tract. The association of ureteral triplication and obstruction is even rarer. We report a rare case of ureteral triplication with hydronephrosis due to ureteropelvic junction (UPJ) obstruction and stenosis at the upper pole ureter. To the best of our knowledge, this is only second such case reported in literature. The child underwent excision of the obstructed segment with a side to side pyeloureterostomy. We salvaged the kidney and the patient is well 20 months postoperatively with no urinary infection and stable renal function. The embryology and management of this rare case along with review of literature is discussed herewith.

INTRODUCTION

Ureteral triplication is a very rare anomaly of the urinary tract. 1, 2, 3–8 Since the first description by Wrany in 1870, only approximately 100 cases have been published till now.⁹ Hydronephrosis due to ureteropelvic junction (UPJ) obstruction in a triplicate system is even rarer. Paucity of symptoms often leads to missed or delayed diagnosis of the entity. Proper diagnosis and timely intervention can save the functioning renal tissue from progressive destruction.

CASE REPORT

A 2 ½ year, male was admitted with the complaints of persistent right abdominal pain and recurrent urinary tract infection, since 4 months. No other relevant positive history or complaints were noted. Clinical examination was unremarkable. Ultrasound (USG) abdomen showed right side hydronephrotic kidney with suspicion of duplex system and left side normal kidney. Micturating cystourethrogram (MCU) showed a urinary bladder of normal outline with no filling defect or pressure effect. No vesicoureteral reflux (VUR) was noted on either side. Intravenous urography (IVU) and Magnetic resonance urography (MRU) revealed the presence of right side duplex system with upper moiety PUJO and hydronephrosis [Figure. 1 and 2]. Cystoscopy revealed single ureteric orifice in normal positions on each side. Armed with the above findings and with a suspicion of duplex system on the right side, the patient was explored with right flank approach in lateral decubitus.

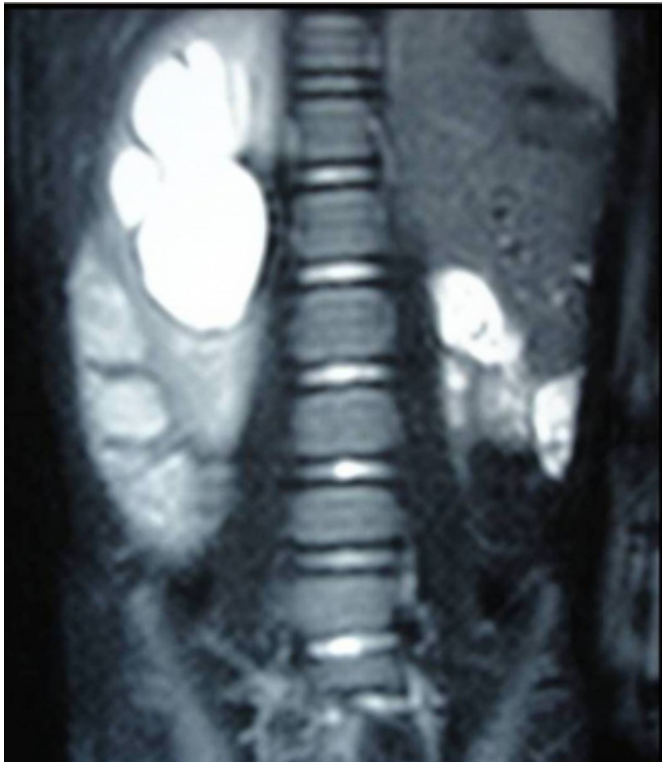
Figure 1

IVU showing presence of right side (?) duplex system with upper moiety PUJO and hydronephrosis



Figure 2

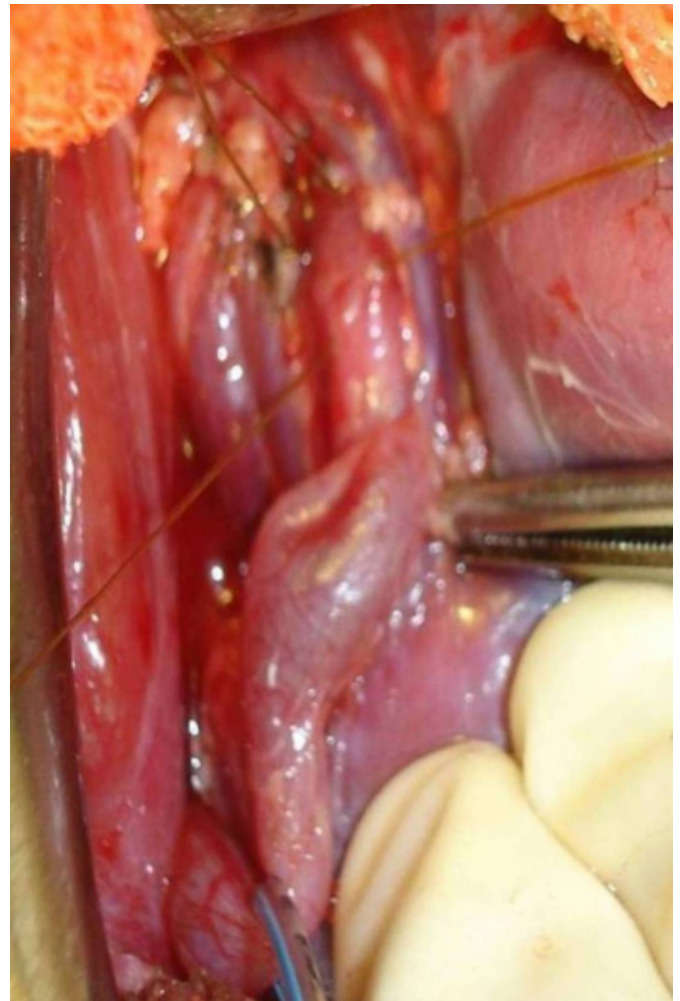
MRU suggestive of right side upper tract anomaly with upper moiety PUJO and hydronephrosis



A huge cystic swelling noted in the right hilar region was obscuring the anatomy, which became clear after aspiration of the cyst. In contrast to preoperative investigations, intra-operative findings showed triplication of renal pelvis with upper moiety PUJO with a thin parenchyma and markedly dilated upper renal pelvis, while middle and lower moieties were normal [Figure 3].

Figure 3

Intra-operative findings showing triplication of renal pelvis



The findings were consistent with type 3 ureteral triplication according to the Smith's classification. The obstructed segment was excised and a side to side pyeloureterostomy was performed over a 3 Fr 16 cm DJ stent [Figure 4a and b]. The patient recovered uneventfully and was discharged on postoperative day 3. Patient got admitted twice subsequently with urinary tract infections, both of which resolved with antibiotics. Postoperative serial USG abdomen showed marked improvement in hydronephrosis. Postoperative nuclear renal scans showed significant improvement in kidney drainage on washout curves confirming decompression and relief of obstruction. Patient is presently asymptomatic at 20 month follow up.

Figure 4a

Diagrammatic representations of intra-operative findings and side-to-side pyeloureterostomy

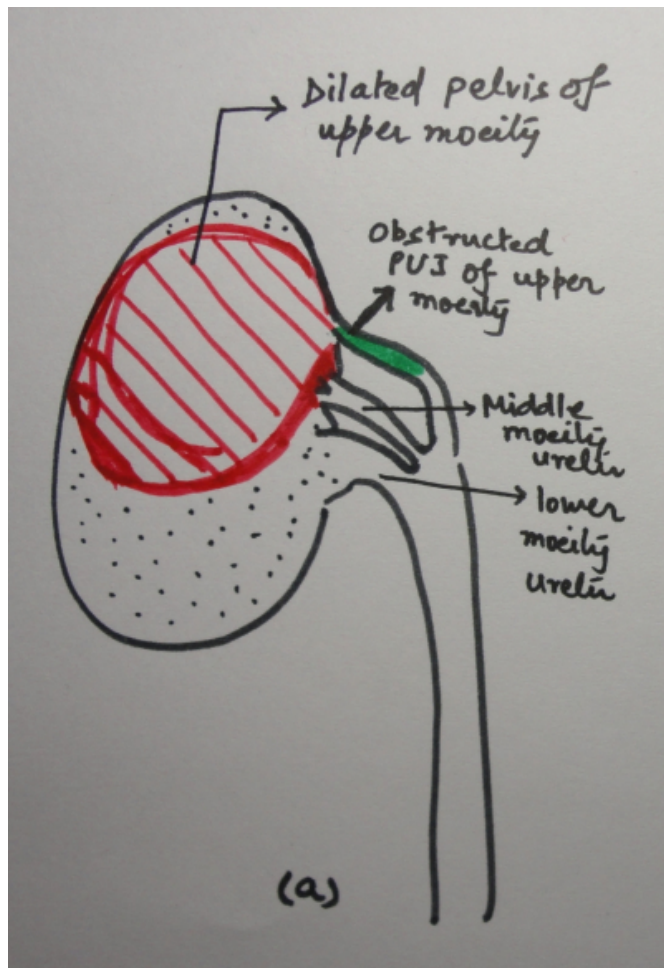
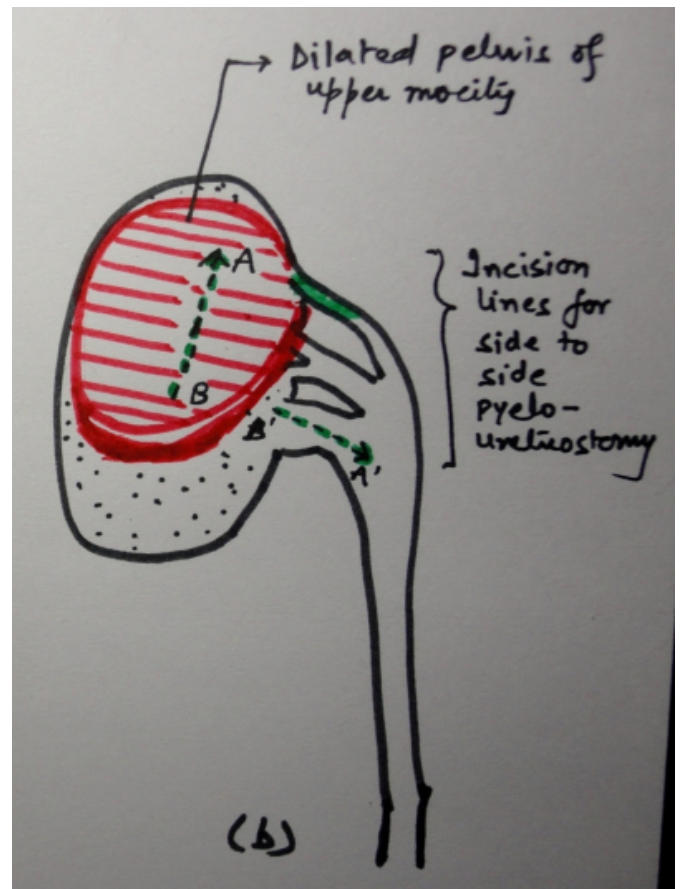


Figure 4b

Diagrammatic representations of intra-operative findings and side-to-side pyeloureterostomy



DISCUSSION

Ureteral triplication is an extremely uncommon congenital urinary anomaly. It is a developmental abnormality of the ureteral bud originating from the Wolffian duct at the 50 week of embryological life. 1 Distal part of the wofflian duct gives rise to the ureteral bud duct after 4 weeks of fetal development. It first grows dorsally and then cranially to make contact with metanephros. During the 6th–8th weeks the distal end differentiates into the renal pelvis and the major and minor calyces. In triplication of the ureter three ureteral buds arise either independently from the mesonephric duct, or result from early fission of one or more ureteral buds while joining the metanephros. 2, 4, 7, 8 Ureteral triplication is more common in females and commonly located at the left side unlike in this case where it was right sided. 2, 5, 7

Smith has classified Ureteral triplication into four types. 1-5, 7, 8

Type 1: Complete ureteral triplication (35%); three separate ureters from the kidney with three separate draining orifices

to the bladder or elsewhere in the urogenital tract.

Type 2: Incomplete triplication (21%); three ureters arise from the kidney but two of these join draining into two ureteric orifices.

Type 3: Trifid ureter (31%); all three ureters join together before reaching the bladder and drain through a single orifice. The case presented herein is classified as type 3.

Type 4: Double ureter one of them bifurcated (9%); two ureters arise from the kidney, one becoming an inverse Y bifurcation, draining into three orifices.

Unlike the duplex systems the positions of the ureteral orifices in triplication of ureter do not always follow the Meyer-Weigert law (the laterocranial ostium corresponds to the caudal renal pelvis and the mediocaudal ostium to the cranial renal pelvis).^{10,11} Out of eight patients with Type-1 ureteral triplication described by Perkins et al five patients did not conform to the law.¹²

The incidence of renal and other congenital anomalies are increased in ureteral triplication. The most frequently encountered urological anomalies associated with ureteral triplication are contralateral duplications (37%), ureteral ectopia (28%) renal dysplasia (8%) and reflux.^{1, 3, 8} Finkel and Gosalbez reported ureteroceles associated with type I and rarely with type II Ureteral triplications.^{3,13}

The most common presenting symptoms in ureteral triplication are: renal colic, recurrent urinary tract infection and urinary incontinence^{1, 5}. Our case also presented with recurrent urinary tract infection and flank pain. For diagnosis USG and MCU although helpful are not very precise for the diagnosis. Dynamic MR nephrography which combines functional and morphologic analyses of the urinary tract has been recently described as additional diagnostic tool for selected pediatric urological cases.^{14,15} It provides a higher spatial resolution compared with sonography and allows a more precise definition of regions-of-interest than renal nuclear scan with additional benefit of not using any ionizing radiation.^{14, 16-18} Additionally, renal scarring and the split renal function can be assessed sufficiently through MR urography which obviates the need for renal scan.¹⁹

Several aspects need detailed evaluation whenever surgery is necessary in triplicate ureters.

Functional evaluation by dynamic MR nephrography yields the required information. It allows precise region-of-interest

evaluation within the renal parenchyma and the urinary passage.^{17, 18} The exact correlation between the preoperative imaging and the intraoperative situs underlines the superiority of the MR nephrography in this regard. In comparison with renal scintigraphy, the higher spatial resolution of the dynamic MR imaging examination and the precise excretion study provided reliable information improving the management of the patient.¹⁹

Magnetic resonance nephrography should be considered as the diagnostic tool of choice for preoperative planning in cases of ureteral triplication. The management of these cases is individual and depends on the clinical presentation, the evidence of obstruction, or reflux

On recent review of the literature, Pode and Merlini reported Ureteral triplication cases associated with obstruction which was caused by uretero-vesical stenosis and ureteroureteric reflux respectively.^{6, 7} In both the cases the obstructed renal segment had to be excised. The only case of ureteral triplication associated with ureteropelvic junction obstruction was reported by Sivrikaya et al which was Smith's type 3 with middle pole UPJO. Our case also had type 3 Ureteral triplication but upper moiety was obstructed with a thin parenchyma and markedly dilated renal pelvis. Similar to our case they were also able to preserve the obstructed renal segment by pyeloplasty.

While many patients are asymptomatic or have only mild symptoms, early diagnosis of ureteral triplication is important to avoid complications and progressive renal damage.²⁰ In our case, diagnosis and timely intervention lead to prevention of any further kidney damage

In conclusion, this case emphasizes one more time the importance of early diagnosis with complete anatomic and functional evaluation of the urinary tract in the management of the patients with recurrent urinary tract infections. Magnetic resonance nephrography should be considered as diagnostic tool of choice for preoperative planning in cases of ureteral triplication. The management of these cases should be individualized depending on the clinical presentation, the evidence of obstruction, reflux and renal function. Early operative therapy can successfully prevent decrease or loss of renal function.

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