

Ureter Quadruplication with Huge Ureteral Cyst

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Abstract

Ureteral quadruplication is an extremely rare congenital anomaly. It was first reported in 1975. The patients may present with different features and complications in respect to this congenital anomaly. They may also present with other congenital anomalies. Diagnosis sometimes is difficult and may need to be investigated with Magnetic Resonance Urogram, CT scan apart from routine investigations like ultrasonography and intravenous urogram. Surgical treatment should be based on the operative and investigational finding. We present the twelfth reported case but the first from Nepal. It was associated with a large ureteral cyst with diagnostic dilemma. Surgical exploration was done with a provisional diagnosis of large retroperitoneal cyst.

Key words: Congenital anomaly, Intravenous Urogram, Quadruplication, Ultrasonography, Ureteral cyst

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Introduction

Ureter quadruplication is the rarest congenital anomaly among ureter malformations^{1,2,3,4}. First reported case was by Sodedahl DW et al in 1976². Since then 11 such cases have been reported worldwide. Our paediatric case is the first to be reported from Nepal and twelfth in the world. This malformation may be associated with vesico-ureteral reflux (VUR)⁵, dystrophic kidney⁶, or blind ending proximal branches⁷. These patients may also present with contra lateral malformation like contra lateral multicystic dysplastic kidney⁸, triplet ureter, agenesis of the kidney or even bilateral ureteral quadruplication². Recently in 2014 quintoplication of ureter has been reported by Jurkiewicz et al which is first of its kind⁹. The present case is of a girl of 5 years old who presented with distended painful abdomen. After relevant investigations we were unable to come to a definite diagnosis for which surgical exploration was carried out.

The Case

A five year old girl child presented with abdominal distension and episodic pain in her right side of the abdomen. The intensity of pain had increased for past 20 days. She had no history of urinary or bowel problems in the past. On physical examination, the patient had distended painful abdomen on the right side. She had no fever,

jaundice, or pallor. Haematological and biochemical studies were all within normal limits. Her urine showed plenty pus cells with 75-80 rbc /hpf and *E. coli* on urine culture. Ultrasonography reported as thick walled retroperitoneal cyst with right mild hydronephrosis and bilateral malrotated kidneys. On Intravenous urogram (IVU), it showed right mild hydronephrosis with bilateral malrotated kidneys (Figure 1). With no obvious conclusion CT-scan was done and revealed thick walled retroperitoneal cyst abutting the right kidney with internal contrast in delayed image and duplex right kidney with obstructed hydronephrotic lower moiety ? Urinoma (Figure 2). She was admitted and intravenous antibiotic

was started as per her culture and sensitivity of urine culture.

After counselling about the difficulty in diagnosis, patient's parents agreed on surgical exploration and also for possible nephrectomy. On exploration, we found a large retroperitoneal cyst which on further dissection kidney was found attached to it with four branches (Figure 3). Not recognizing what they really were, we cut two branches thinking it as adhesion. On due course of dissection, we noticed a long tube distally connected to the cyst. We were unable to understand the anomaly, we requested our one of the resident to web-search immediately. We later concluded this must be a case of quadruplication of ureter with large ureteral cyst and a single ureter (Figure 4). Kidney looked grossly normal as IVU and CT-scan suggested, functioning kidney though DTPA scan was not done. The iatrogenic injury of two upper moiety were anastomosed by Wallace fashion and then to the cyst. The large ureteral cyst underwent reduction pyeloplasty with the single ureter distally as there was a partial stenosis of about 1 cm at the junction of cyst and ureter. DJ stent of 5 French size was placed to safeguard the anastomosis. The girl was hospitalized for 7 days with removal of drain on the 6th day post-operatively. On follow up after 6 weeks she was free of any post-operative complication and DJ stent was removed. Her parents have been advised to attend the follow up clinic on 3rd and 6th month. She will undergo IVU and possibly DTPA scan on the 6th month.



Fig 1: Intravenous urogram showing malposition of Kidneys

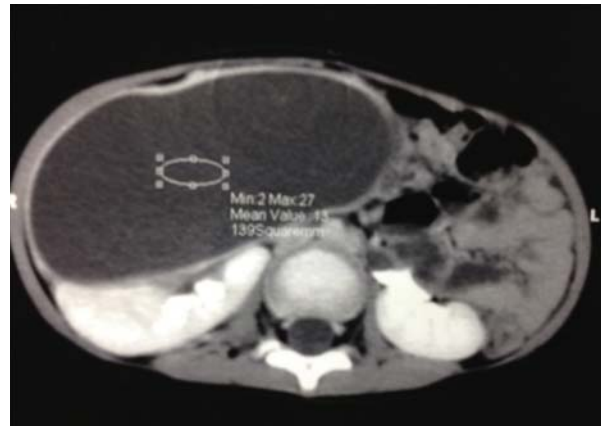


Fig 2: CT scan showing large retroperitoneal cyst with functioning kidney

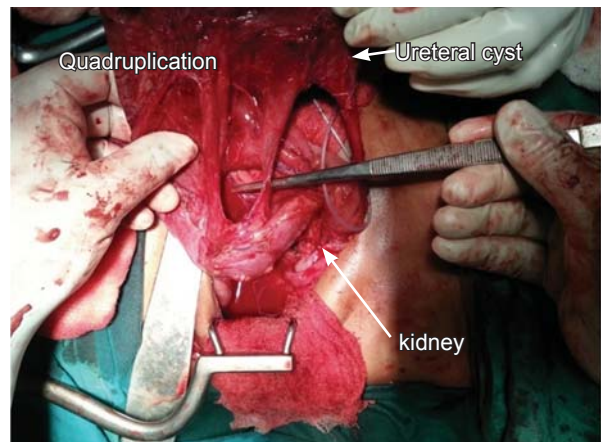


Fig 3: Showing quadruplication of ureter, kidney, large ureteral cyst and single ureter

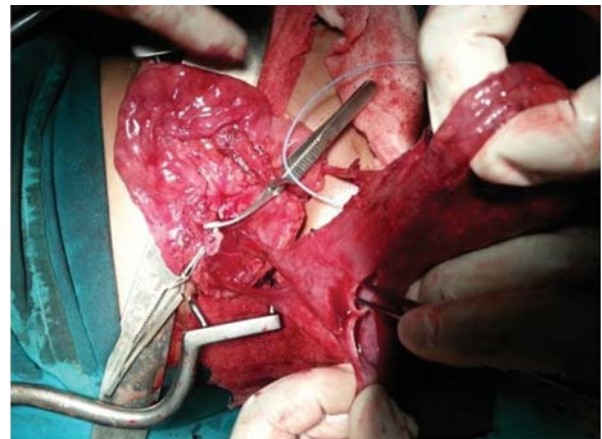


Fig 4: Showing Cut ends of upper moiety, lower moiety, kidney, ureter, ureteral cyst

Discussion

During the early fourth week of gestation ureteric bud arises as a diverticulum from the Wolffian duct. Once in contact with metanephric mesenchyme it undergoes a dichotomous branching morphogenesis¹⁰. It has been postulated that AT2 (angiotensin II receptor)

plays a vital role in directing the site of ureteric bud outgrowth. The defect in this process leads to defective timing and location of outgrowth of ureteric bud leading to congenital ureteral anomalies¹⁰.

Quadruplication of the ureter is a very rare type of congenital anomaly. The first reported case was by Soderdahl in 1976 and only 11 such cases has been reported worldwide till date². Our case is unique as it presented with huge distended abdomen due huge dilatation of ureteral cyst caused by stenosis of “ureterocysto-pelvic junction” with normal four congenital ureter. Triplication of ureter as described in literature has four varieties according to Smith classification but no such variety is described for the quadruplication of ureter. Quadruplication is associated with many other anomalies like three blind-ending braches, multicystic dysplastic kidney, bilaterality and massive vesicoureteral reflux in the contra-lateral side.

Diagnosis is usually made during surgical exploration as in our case. Preoperative evaluation with IVU, retrograde pyelography may show congenital abnormalities. CT-scan and VCCUG may be needed for further evaluation to look for other ipsi /contralateral anomalies and VUR. DMSA or DTPA scan will indicate the functional status. Magnetic Resonance urography (MRU), if available, may be more diagnostic and may provide high quality images of the urinary tract without using ionizing radiation. MRU can provide both excretory functional and morphological information by using diuretic and contrast media.

There are no general consensus on the surgical procedure .It may differ due to varied presentation of the anomaly. Our case underwent Wallace anastomosis for the iatrogenic injury of the upper two moiety and reduction pyeloplasty for “ureterocysto-pelvic stenosis”. The kidney looked healthy though DTPA scan was not done preoperatively. Massive VUR with impaired renal function can undergo nephroureterectomy⁵. Akaba et al¹ preserved the kidney by termino-terminal pyelorenal anastomosis. To preserve the afflicted kidney is justifiable and defendable as in our case but the patient needs to be in regular follow up. Our patient is still under routine regular medical follow up. At sixth month the patient will undergo test like IVU and DTPA to look for post-operative complications like re-stenosis and functional condition.

Conclusion

Quadruplication of ureter like our case is an extremely rare ureter congenital anomaly. Very few cases are reported worldwide and there is no consensus regarding its treatment options

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