ORIGINAL RESEARCH PAPER

Urology

QUADRUPLE URETER IN A DUPLICATED SYSTEM WITH NON FUNCTIONING UPPER MOEITY- A RARE CASE REPORT

KEY WORDS: Congenital anomaly, Intravenous Urogram, CT scan, Quadruplication, Duplicated moiety

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ABSTRACT

Ureteric quadruplication is an extremely rare congenital anomaly. It was first reported in 1975. They are usually associated with other congenital anomalies and usually symptomatic. Surgical treatment varies and is based on investigational and operative findings. Our patient was a middle aged woman presented with left flank pain. We present radiologic and cystoscopic images of this patient .CT scan revealed left duplicated moiety with non functioning upper moiety. Cystoscopy showed ectopic 3 ureteric orifices just distal to bladder neck along with bilateral normal orifices. She underwent left heminephrectomy.

Introduction

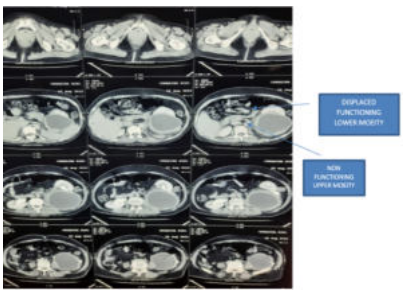
Genito urinary malformations comprise about 3% of live births.[1] Ureters are a pair of thick-walled cylindrical tubes, each measuring about 25 cm in length and 3 mm in average diameter, extending from the funnel-shaped renal pelvis to urinary bladder (UB), and convey urine from the corresponding kidney.Ureter develops from stalk of ureteric diverticulum which grows cranially from the caudal part of mesonephric duct around the 5th week of intrauterine life, that lies between the renal pelvis and vesico urethral canal.[2] Anatomical variations of ureter and its relationship to surrounding structures are therefore important in academic as well as clinical perspective to preserve renal functions. Around 14 cases od quadruple ureter have been reported till now making it a very rare upper urinary tract abnormality.[3]

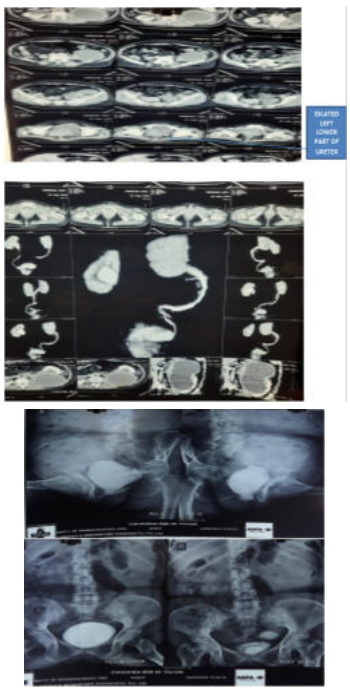
Case presentation

40-year-old non-pregnant woman presented with right flank pain of 6 months' duration. Pain was insidious in onset, gradually progressive, dull aching in nature, no aggravating factors, no radiation, relieved with medication. She was a known hypothyroid since 10 years well controlled on thyroxine(50).

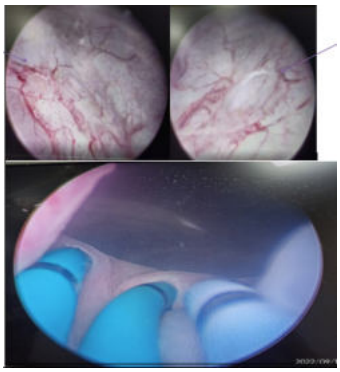
She had no additional symptoms or significant past history. There were no physical findings, and urine examination, renal function tests and serum biochemistry were normal. Ultrasound revealed left severe hydronephrosis.

CT scan revealed left duplicated moiety with severely dilated sac like upper moiety with no discernible uptake of contrast and compression of normal functioning lower moiety upwards and laterally, along with dilatation of lower part of ureter into urinary bladder (? Ureterocle)

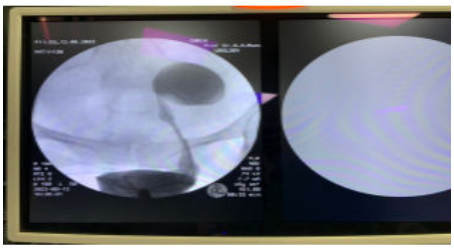


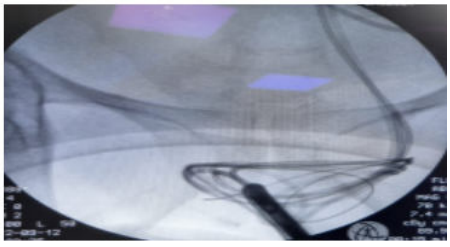


MCU SHOWING DILATATION OF LOWER URETER AREA IN REGION OF BLADDER (? URETEROCELE) WITH NO EVIDENCE OF VUR



CYSTOSCOPY S/O BILATERAL NORMAL URETERIC ORIFICES;
DISTAL TO BLADDER NECK- 3 ECTOPIC URETERIC OPENINGS SEEN - AND CANNULATED
NO URETEROCELE NOTED;
GUIDEWIRE FROM 3 ECTOPIC ORIFICES CONNECTING AT ONE PLACE



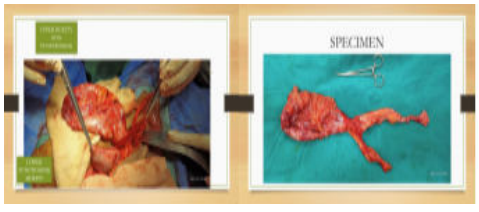


RGP FROM ALL 3 ECTOPIC ORIFICES SHOWING COMMON DYE PASSAGE INTO SAC LIKE UPPER MOEITY.

RGP FROM LEFT ORTHOTOPIC ORIFICE SHOWING DYE PASSAGE INTO LATERALLY DISPLACED LOWER MOEITY

Based on radiologic and cystoscopy findings, a diagnosis of left sided double moiety system was made, with upper non functioning moiety compressing lower moiety, upper moiety having 3 ureters with 3 ectopic orifices just distal to bladder neck and lower moiety having one ureter with normal opening.

Our patient underwent open left heminephroureterectomy of upper moiety with ureter being ligated as distally far as possible, taking care not to damage the lower moiety kidney and ureter.



Discussion

During the early fourth week of gestation, one ureteric bud arises as a diverticulum from the Wolffian duct. Once in contact with metanephric mesenchyme it undergoes a dichotomous branching morphogenesis.

It has been postulated that Angiotensin II via AT2 receptor plays an important role in directing the site of ureteric bud outgrowth. If there is any defect in this process, it leads to defective timing and location of outgrowth of ureteric bud resulting in congenital ureteral anomalies[4]

Triplication of ureter as described in literature has four varies according to Smith classification but no such variety is described for the quadruplication of ureter.

This malformation may be associated with vesico-ureteral reflux (VUR)[5] , dystrophic kidney[6] , or blind ending proximal branches[7] . These patients may also present with contralateral malformation like contralateral multicystic dysplastic kidney[8] , triplet ureter, agenesis of the kidney or even bilateral ureteral quadruplication.[9]

Quadruplication is associated with many other anomalies like three blind-ending braches, multicystic dysplastic kidney, bilaterality and massive vesicoureteral reflux in the contralateral side.

Diagnosis is usually made during pre operative imaging, cystoscopy or ultimately during surgical exploration as in our case. Magnetic Resonance urography (MRU), if available, may be more diagnostic and may provide high quality images of the urinary tract without using ionizing radiation.

There is no definite surgical intervention, as presentation varies in different patients with quadruple ureter and ultimate decision to be taken at the time of surgery.

Conclusion

Very few cases of quadruple ureter are reported worldwide. Quadruplication of ureter is a rare ureter anomaly, and its association with duplicated system with a non functioning upper moeity like our case is an extremely rare presentation. The presentation varies in different patients and there is no general consensus regarding its treatment option.

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