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Urology

WEIGERT MEYER LAW – COMPLETE DUPLEX KIDNEY & URETER WITH OPENING OF ECTOPIC URETERIC ORIFICE AT THE BLADDER NECK IN ADULT INDIAN FEMALE

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ABSTRACT A duplex kidney is characterized by the presence of two separate pelvicalyceal systems with complete or partial duplication of the ureters. Duplex kidneys if causing symptoms usually present in childhood, rarely present in adults. Presenting a rare case report of a 37 years female presented with right flank pain with fever with vomiting and dysuria complain. Past h/o 2 LSCS. 1st time diagnosed as right side gross hydronephrosis of upper pole with complete duplex collecting system with duplex ureter with opening of ectopic ureter at bladder neck and non-functioning upper moiety. She underwent laparoscopic converted to open partial Nephro-uretrectomy due to severe adhesions and bleeding. Post-operative course was uneventful. In conclusion, a thorough evaluation with a high index of suspicion is required in individuals presenting with flank pains to quickly identify hidden duplex renal systems and related complications.

KEYWORDS : weigert meyer law, complete duplex ureter, adult Indian female

INTRODUCTION –

Ureteral duplications are a common congenital anomaly of the kidney and the urinary tract system [1, 2]. It has an incidence rate of 0.8% in the healthy adult population, 0.3%–2.5% incidence in an autopsy, and 2–4% in patients investigated for urinary tract symptoms [3]. Complete duplication occurs in 40% of cases, whereas partial duplication occurs in 60% of the duplication [3]. The Weigert- Meyer law is applicable to a completely duplicated collecting system. It states that the ureter arising from the lower pole moiety runs a short intramural course and inserts into the urinary bladder more superiorly and laterally. This predisposes the ureter to reflux [4,5]. The other ureter arising from the upper pole of the kidney inserts more inferiorly and medially and is prone for ureterocele and obstruction. Most patients remain asymptomatic despite the relatively common incidence. Patients usually present in childhood, however, in rare instances can present as adults [6]. To the best of our knowledge, only a few cases are reported of a patient with unilateral complete ureteral duplication with ectopic uretric opening at bladder neck. We present a case of a hydronephrotic upper moiety of the right complete duplex system with hydro-ureter with ectopic uretric opening at bladder neck in adult Indian female.

Case Report –

Patient Information -

A 36-year-old female patient presented with complain of right flank pain with fever with dysuria. The right flank pain was present for 3 months and it was a dull aching type, not radiating and there were no aggravating or relieving factors. The fever and dysuria were present for last 3 days. There was past history of previous 2 LSCS.

Clinical Findings -

On physical examination, her vital signs were normal. In the abdominal examination, tenderness in right hypochondrium with right renal angle tenderness was present. Previous lower section caesarean scar was present.

Diagnostic Assessment -

Blood tests revealed a total white blood cell count of 5.5 × 10^3/ml, haemoglobin of 9 gm/dl, blood urea nitrogen of 14 mg/dl, and creatinine of 1 mg/ dl. Ultrasound KUB showed right kidney upper pole gross hydronephrosis with hydroureter present, lower pole of right kidney was normal. There was duplication of right renal collecting system and ureter. CT urography showed normal excretion of the left kidney and ureter with no obstruction. Right kidney showed similar changes as mentioned above (figure 1 & 2). DTPA renal scan showed non-functioning of right upper pole moiety with suboptimal functioning non-obstructed right lower pole moiety. Normal functioning, non-obstructed left kidney (figure 3).

Therapeutic Interventions -

Injectable antibiotic (meropenem, 1 gm, TDS) and symptomatic treatment was started on admission. On basis of investigations, patient was planned for Laparoscopic partial nephrectomy of hydronephrotic non-functioning upper pole moiety with uretrectomy. Under general anaesthesia, in lithotomy position cystoscopy was done, which showed Ectopic ureter opening at bladder neck with wide-mouth (figure 4). Modified left lateral decubitus position given, standard laparoscopic port placement done. Ascending colon mobilized was from white line of toldt and right side kocherization was done. Retroperitoneal space was entered and right kidney was identified after Gerota's fascia was opened. Kidney was densely adherent to surrounding structures and with IVC. Due to severe adhesions and bleeding laparoscopy converted to open partial nephroureterectomy. Approx 350 cc pus came from right upper moiety and 200 cc pus came from ectopic ureter. Both the ureters were in common sheath till the upper border of bladder, upper third of ectopic ureter transacted and ligated just above the upper border of bladder. Some Pus spillage occurred in peritoneal cavity, through peritoneal wash was given. An intra-abdominal drain was left in situ, and the abdominal wall was closed in layers.

Follow-Up and Outcome -

The patient had a smooth postoperative course following the surgery. The drainage tube was removed after 36 hours postoperatively. The patient was discharged on her fifth postoperative day. The patient had uncomplicated post operative recovery.

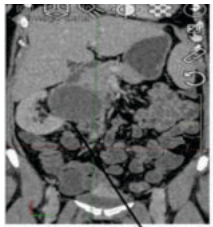

Figure – 1

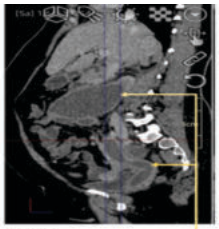

Figure -2

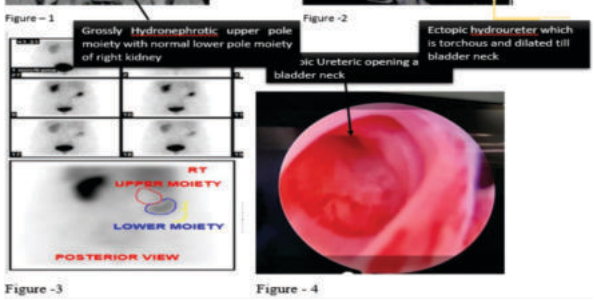

Figure -3

Figure - 4

DISCUSSION –

A duplex kidney is characterized by the presence of two separate pelvicalyceal systems with complete or partial duplication of the ureters, this condition is a known congenital anomaly of the urinary tract [1,2]. According to the Weigert Meyer law, the orifices of the ureters draining the upper pole of the kidney open inferior and medial to the orifice draining the lower pole of the kidney. This was seen in 85% of subjects with complete duplications of ureter [5].

During embryogenesis and development, formation of 2 ureteral buds followed by failure of contact of one of the buds with the metanephrogenic blastema results in the double ureteral orifice formation. This ureteral duplication may be complete or incomplete and is genetically determined by an autosomal dominant trait with incomplete penetrance [5]. At about 4-6 weeks of gestation, the distal mesonephric duct gets absorbed into the urogenital sinus which results in the separation of the ureteral bud and the mesonephric duct orifices [4]. It is reported that this condition is seen in females more commonly with the female to male ratio of 6:1 [4]. In such cases of duplex kidney with double ureter, the lower moiety is most commonly affected by vesico-ureteral obstruction and reflux, whereas the upper moiety is most commonly affected by hydronephrosis. The same finding was observed in our case. It is important to know whether the ectopic ureter shows reflux before planning a partial nephrectomy since it is difficult to dissect the ureter distally if it is inserting beyond the internal ureteral sphincter.[2] In our case, the ureter of the upper renal moiety appeared dilated while the ureter of the lower renal moiety appeared normal.

Most patients remain asymptomatic despite the relatively common incidence. Patients usually present in childhood, however, in rare instances they can present as adults. Presentation can include recurrent urinary tract infections, flank pain, incontinence and haematuria [6]. Imaging modalities used include ultrasound, nuclear medicine, excretory urogram and CT. In adults, CT often demonstrates hydronephrosis, with a greater prevalence in the upper pole moiety. Nuclear Imaging is required for functioning assessment [7].

When symptomatic, the recommended treatment for a duplex system is partial nephrectomy of non-functioning part with uretrectomy. Surgical approaches can be either open or laparoscopic, with laparoscopic gaining more favour but in complicated cases be prepared to convert open such as in our patient.

CONCLUSION –

Duplex kidneys are usually clinically silent. When they are symptomatic, tend to present in childhood. In rare instances, duplex kidneys can appear in adults, leading to diagnostic challenges. The imaging modality of choice is CT, but Nuclear Imaging is required for functional assessment. A thorough evaluation with a high index of suspicion is required in individuals presenting with flank pains to quickly identify hidden duplex renal systems and related complications and plan surgery of partial nephro-uretrectomy and its related complications.

Abbreviations –

KUB – kidney - ureter – bladder
CT – computed tomography
DTPA – diethylenetriamine pentaacetate
LSCS – lower segment caesarean section
IVC – inferior vena cava
TDS – ter die sumendus (three time in a day)

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