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RETROCAVAL URETER: A CASE SERIES OF 5 CASES SUCCESSFULLY MANAGED BY URETEROURETERAL ANASTOMOSIS.

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Abstract

Retrocaval ureter also known as pre-ureteral vena cava is a rare congenital anomaly as ureter passes posterior to the inferior vena cava. It is a congenital anomaly but patients do not present with symptoms until the 3rd and 4th decades of life. Incidence is about 1 in 1500. M:F ratio is 3:1 male predominance in autopsy studies [1]. Complications of retrocaval ureter are urinary tract infection, hematuria. In the present series, 5 case diagnosed with retrocaval ureter were analyzed for age, sex, laterality, symptoms, and degree of Hydronephrosis. Open ureteroureterostomy with double J stent insertion was performed on all 5 patients. Double J stents were removed after 3 weeks and patients were followed at 3 months.

Key words : Retrocaval ureter (RCU), Inferior vena cava (IVC), Hydroureteronephrosis, Uretero-uretral Anastomosis.

Introduction

Retrocaval ureter (RCU) is a rare congenital anomaly in which the ureter passes posterior to the inferior vena cava (IVC). [2] This anomaly occurs between the 4th and 8th weeks of intrauterine development. It is due to abnormal formation of infrarenal IVC from anteriorly located subcardinal vein instead of supracardinal vein which are located posteriorly. In normal circumstances, the infrarenal IVC originates from dorsally located supracardinal vein, but when it develops from ventrally located subcardinal vein, the ureter is trapped posteriorly leading to pre-ureteral vena cava. [3] It was first described in 1893 by Hochstetter in a cadaver. [4] The first clinical diagnosis in 1940 by Harrill. [5] RCU has an incidence of 0.06–0.17% worldwide.
[6] We present five cases of retrocaval ureter which were successfully managed.

Case series:

CASE 1

27 years male came to surgery OPD with history of pain in abdomen, loss of appetite, constipation, mild fever and burning during micturition. USG was suggestive of right 17x10 mm size mid ureteric calculus causing hydronephrosis and Hydroureter. CT scan suggestive of right retrocaval ureter with proximal hydroureteronephrosis. Patient underwent surgery by a right subcostal lumbotomy incision. On exploration, revealed the retrocaval passage of the ureter with a moderate dilatation of the pelvis. Uretero-ureteral anastomosis was performed by four points of Vicryl 3/0 around a JJ stent, then a closure was performed with placement of the drain. Removal of the ureteral catheter on the 6th postoperative day.

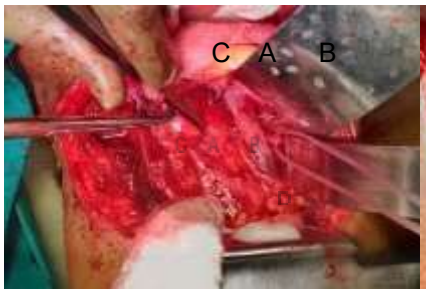
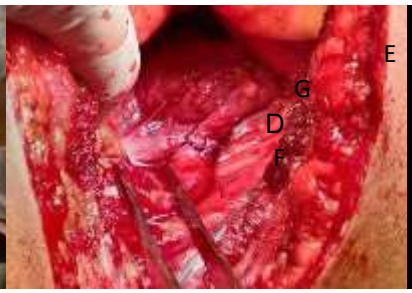


Fig: 1



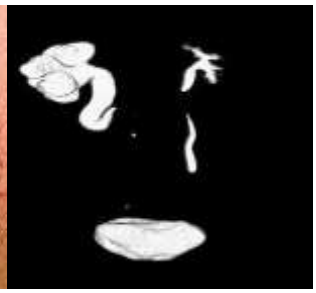


Fig:3 CT Abdomen Pelvis A: Aorta.

B: Inferior vena cava. 1] Right retrocaval ureter
C: proximal portion of the ureter. with proximal
D: distal portion of the ureter. hydroureteronephrosis.
Fig: 2 2] Fish hook appearance at level of L4 vertebral body
E: proximal portion of the ureter.
F : distal portion of the ureter.
G:Final appearance after ureteral resection anastomosis.

CASE-2

A 23-year-old woman presented with a history of dull right flank pain of 2 years duration, burning micturition. She was otherwise well and clinical examination of the abdomen was normal. Laboratory evaluation was within normal limits. Abdominal ultrasound showed right hydronephrosis and 5mm right lower ureteric calculus. CT KUB showed right hydroureteronephrosis with an "S" shaped or "fish hook" deformity of the proximal ureter, which terminated abruptly. A diagnosis of retrocaval ureter was made and the findings at operation were that of right retrocaval ureter, proximal dilated ureteral segment and a normal distal segment lying between the aorta and IVC. The redundant retrocaval segment was mobilized and excised, and end-to-end uretero-ureteral anastomosis was achieved over a JJ stent.

CASE-3

A 25-year-old male presented with a history of dull intermittent right flank pain of 1 year duration. He had history of fever, burning micturition, no history of dysuria, haematuria, or weight loss. Laboratory evaluation was normal. Abdominal ultrasound showed 18x16mm right mid ureteric calculus with right hydronephrosis with proximally dilated ureter. A CT urogram showed retrocaval ureter with proximally dilated ureter which was confirmed at surgery. Excision of the retrocaval segment with end-to-end ureteral anastomosis over a stent was done.

CASE-4

A 23-year-old man presented with a history of abdomen pain since 1 year. He had history of vomiting, fever, burning micturition. Laboratory evaluation was normal. Abdominal ultrasound showed 9.2x7mm right mid ureteric calculus with right hydronephrosis. CT-KUB showed Right side retrocaval ureter with S deformity at level of l3 vertebra. Excision of the retrocaval segment with end-to-end ureteral anastomosis over a stent was done.

CASE-5

A 17-year-old female presented with a history of abdominal pain 1 year duration. He had history of fever, burning micturition, haematuria. Laboratory evaluation was normal. Abdominal ultrasound showed 8.8mm right lower ureteric calculus with right hydronephrosis with proximally dilated ureter. A CT-KUB showed retrocaval ureter. Excision of the retrocaval segment with end-to-end ureteral anastomosis over a stent was done.

PARAMETER S	Pt 1	Pt2	PT3	PT4		PT5
AGE(years)	27	23	45	23		17
SEX	Male	Female	MALE	Male		Female
SYMTOMS						
Abdominal pain	+	+	+	+		+
Vomiting and nausea	+	+	+	+		+
Fever	+	+	-	+		+
Hematuria	-	-	-	-		+
Burning micturition	+	+	+	+		+
Investigation						

USG abdomen- pelvis	-17*10mm right ureteric calculus, Moderate hydroureteronephro sis	-5mm right lower ureteric calculus Moderate Right side hydrouretro -nephrosis	-18*16mm right mid ureteric calculus Moderate Right side hydrouretro -nephrosis	-9.2*7 mm right mid ureteric calculus mild Right side hydrouretro -nephrosis	-8.8mm right lower ureteric calculus Moderate Right side hydrouretro -nephrosis
CT scan	Right Retrocaval ureter at level of L3 vertebra	Right Retrocaval ureter at level of L3 vertebra	Right retrocaval ureter at level of L3 vertebra	Right Retrocaval ureter at L3 vertebra	Right Retrocaval ureter at L3 vertebra
Associated congenital anomaly	No	No	No	No	No
Treatment • **Conservative** • **Surgical**	Uretero-uretral Anastomosis	Ureterouretral Anastomosis	Ureterouretral Anastomosis	Ureterouretral Anastomosis	Ureterouretral Anastomosis

Discussion

Retrocaval (circumcavel or postcavel) is a rare congenital abnormality. Retrocaval ureter is a developmental abnormality of the venous system. Inferior vena cava is made from three bilateral venous systems. The posterior cardinal veins, longitudinal in the dorsolateral position, the subcardinal veins in the medial position and the supracardial veins responsible for the final formation of the inferior vena cava. The persistence of the posterior cardinal vein is at the origin of the retrocaval ureter. [7] The majority of retrocaval ureters presents with various clinical manifestations, which are related to upper urinary tract obstruction and its complications. [8] CT scan is helpful in identifying malformations like pyelo ureteral dilatation, intervertebro-cave passage of the ureter and then its anterior pre-cave path. Using the CT scan, it is possible to make a differential diagnosis with primary idiopathic or secondary retroperitoneal fibrosis, and with any other cause of ureteral obstruction. [9] An anatomical classification, proposed by Kenawi and Williamsen in 1976.[10] Distinguishes retrocaval ureter into two anatomical types according to the height of the retrocaval segment of the ureter. This is based on the radiographic appearance and the location of the ureter's narrowing. Type 1 is most common anomaly and occurs in 94% of cases. The path of the ureter is normal up to height L3, where it then passes behind the inferior vena cava. Type 2, the pelvis and the initial segment of the ureter occupy an almost horizontal position, whereas in type 1 the curve which the ureter forms when passing behind the inferior vena cava is slight, so that the degree of dilatation of the pyelocaliceal system and the ureter is less pronounced in this type. A classification of surgical interest has been adopted by Bateson and Atkinson, [11] who consider that the obstruction mechanism is different for the two types of retrocaval ureter: Type 1 in which the obstructive syndrome is due to a intrinsic anomaly in the development of the retrocaval segment of the ureter requiring surgical resection, type 2 in which the obstruction is due to extrinsic compression of a normal ureter in its retrocaval portion. The treatment depends on the degree of obstruction of the malformation and the radio clinical follow-up. The nephrectomy is necessary in the rare cases of destroyed kidneys. Between these two extreme situations, the place of conservative surgery is preponderant and constitutes the treatment of symptomatic forms. The technique usually used is ureteral uncrossing with restoration of continuity of the excretory pathway by direct plasty and end-to-end Ureteroureteral anastomosis in type 2, and resection-anastomosis of the retrocaval segment in type 1. Laparoscopic reconstructive techniques have been described by several authors who believe that it should be a technique of choice in the surgical treatment of the retrocaval ureter, this technique offers several advantages over conventional open surgery: it is minimally invasive, decreased blood loss, decreased urine leak, shorter hospital stay and recovery time were reported with the retroperitoneal laparoscopic approach compared to open approach allows early emergence, absence of complications such as postoperative wall pain or gastrointestinal ileus, and a shorter recovery period. However, significantly, higher operative time was detected with the laparoscopic repair of RCU. Robotic technology could decrease the operative time during the surgery. The robotic trans peritoneal approach may shorten the operative time enabling an easy surgical dissection and suturing with a greater comfort to the surgeon in the repair of RCU. [12]

Conclusion

Retrocaval ureter is a rare congenital anomaly that presents clinically late in the third and fourth decades of life with equal Male to female preponderance and rarely associated with any congenital anomaly. The diagnosis is confirmed by the Ultrasonography and CT KUB. Surgical treatment of symptomatic cases successfully relieves symptoms.

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