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Case Report

Surgery Section

Ureteral Entanglement in Ring Inferior Venacava- A Rare Presentation

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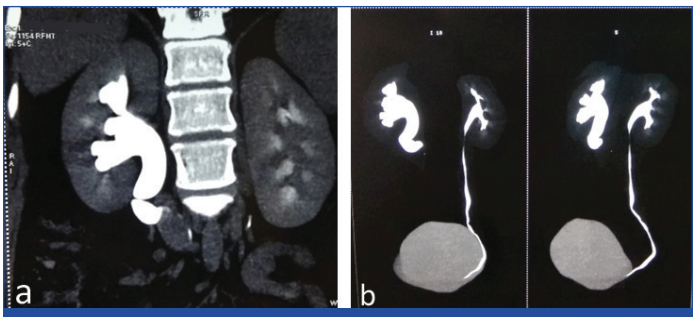
ABSTRACT

Ureteral entanglement in ring Inferior Venacava (IVC) also called Periureteric venous ring is a rare developmental anomaly, where right ureter passes through a slit like opening in a partially duplicated infrarenal IVC, this results in a dilated upper urinary tract. Authors hereby report a case of a 25-year-old married female presented with flank pain of right side of abdomen for seven months. On Contrast Enhanced Computed Tomography of Kidney, Ureters and Bladder (CECT KUB) with urography diagnosed as retrocaval ureter with right hydronephrosis. The patient was planned for right ureteroureterostomy. During surgery, it was found that right ureter was entangled in between slit like opening of inferior venacaval ring. Preureteral venacava with ring IVC is a rare phenomenon. Clinically it is very difficult to distinguish between the causes of ureteric obstruction. There is scarcity of such cases in the literature. The index case was also diagnosed intraoperatively and managed successfully.

Keywords: Inferior venacava duplication, Periureteric venous ring, Upper tract dilation

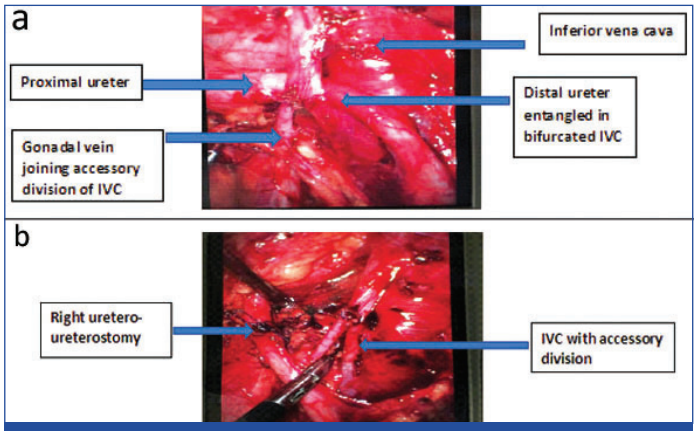
CASE REPORT

A 25-year-old married nulliparous female presented with insidious onset, intermittent, dull flank pain of right side of abdomen for seven months. She had no other co-morbidities. On physical examination abdomen was soft, non tender. No abnormalities were detected in cardiological, neurological and respiratory system examination. Ultrasonography of abdomen showed right hydronephrosis with dilated right ureter. Intravenous urography revealed dilated pelvicalyceal system and proximal ureter on right side. Left side was normal. Contrast Enhanced Computed Tomography (CECT) with urography showed right hydronephrosis, ureter dilated in proximal with narrowing in mid segment [Table/Fig-1a,b], suggesting retrocaval ureter.



[Table/Fig-1a,b]: Contrast Enhanced Computed Tomography (CECT) with urography showing right hydronephrosis, ureter dilated in proximal part with narrowing in mid segment.

Patient was planned for laparoscopic ureteroureterostomy. On preanaesthetic evaluation, no co-morbidities were noted and patient was fit for surgery. At first, ureteric catheter was placed in right ureter by cystoscopy which aided in identification of ureter. On exploration, right ureter was found to compress between IVC and accessory division of IVC, right gonadal vein appeared to join the accessory division of IVC [Table/Fig-2a]. Surgical treatment was directed toward relieving the obstruction and correcting the underlying abnormality. Reposition end to end ureteroureterostomy was done over Double J-stent (DJ) stent [Table/Fig-2b] [1]. DJ stent was removed after four weeks of surgery. She was followed-up postoperatively after six months and she is doing well.



[Table/Fig-2]: a) Right ureter was found to compressed between Inferior Venacava (IVC) and accessory division of IVC, right gonadal vein appears to join the accessory division of IVC. b) Reposition end-to-end ureteroureterostomy was done over DJ stent.

DISCUSSION

Embryological development of IVC is very complex, leading to various congenital anomalies. Preureteral venacava is itself a rare congenital developmental anomaly affecting one in 1500 [2]. In obstruction due to periureteric venous ring, the deviated portion of ureter in effect splits venacava and therefore need not appear medial to pedicles of L3 or L4 as in usual circumcaval ureter [2-4].

The infrahepatic IVC develops from a set of three paired veins appearing between four and eight weeks of life, namely the posterior cardinal, subcardinal and supracardinal veins. Posterior cardinal and supracardinal vein lies dorsally, while subcardinal vein lies ventrally. The internal spermatic vein formed by the subcardinal vein. The definitive right-sided IVC is formed from right supracardinal vein. If the subcardinal vein in lumbar portion fails to atrophy and becomes the primary right-sided vein, the ureter is trapped dorsal to it [3]. A double right vena cava is formed because of the persistence of both the right subcardinal vein ventrally, when the definitive vena cava forms normally and the ventral portion of the primitive ring also persists. The right ureter traps between its limbs by this double vena cava [3].

Variations in IVC are often diagnosed incidentally when radiographs taken for any other cause and these variations may have significant clinical implications [4]. In majority of patient symptoms are owing

to ureteral obstruction and resultant hydronephrosis. Pain may resemble renal colic but can be intermittent, dull aching. Sometimes may present with hematuria. Many a times the resultant silent hydronephrosis is unmasked by unrelated clinical events. Though the presentation is similar to any other cause of hydronephrosis, radiological investigation also fails sometimes to diagnose such rare anomaly [5].

There were very few cases reported as periureteric venous ring. Dillon EH and Camputaro C reported the similar case where they used 3D reconstruction CT for diagnosis [6]. In one case reported by Dillon B and Goodman TR, child with flank pain was diagnosed having periureteric venous ring using MRI [7]. Naik S et al., reported a case where they used Multidetector CT (MDCT) with administration of contrast, for diagnosis of this periureteric venous ring [8].

The differential diagnoses for this venous abnormality include right double IVC, IVC thrombosis, dilated retroperitoneal vessels in portal hypertension, retroperitoneal hypervascular tumors, retroperitoneal lymphadenopathy. Preoperative awareness is important to prevent serious complications during surgery [9].

A case was reported where a patient presented with renal calculi and transitional cell carcinoma on the right side. Periureteric venous ring was diagnosed on preoperative imaging and confirmed on surgical exploration for right radical nephroureterectomy [10]. Rabley A et al., reported a case of asymptomatic obstructive ureterolithiasis due to periureteral venous ring, where they managed the case by preoperative ureteral stenting and retrograde flexible ureteroscopy [11].

Management of periureteric venous ring is based on the clinical presentation. Treatment may not be required for asymptomatic patients. Usually surgical management are required in obstructive uropathy patient, which may involves resection and reanastomosis of the ureter with its distal remainder [10-12].

CONCLUSION(S)

Periureteric inferior venacava venous ring causing upper urinary tract dilation is a rare presentation. Vascular cause of ureteric obstruction should be considered as differential diagnosis even in minimally deviated obstructed ureter. While considering the cases of retrocaval ureter, one should keep possibility of this presentation and should be ready for intraoperative challenges.

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