Circumcaval Ureter—the Paradigm Shift in Diagnosis and Management

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Abstract An anomalous development of infra-renal inferior vena cava leads to circumcaval ureter or preureteral vena cava, a rare congenital anomaly with an autopsy incidence of 0.9 per 1000. We present a case of circumcaval ureter and review the literature with a special emphasis on the paradigm shift seen during the last decade, in diagnostic and management strategies.

Keywords Anatomy · Inferior vena cava · Anomalies · Circumcaval ureter · Preureteral vena cava · Retrocaval ureter

Introduction

Circumcaval ureter first described by Hochstetter in 1893 is an uncommon congenital anomaly, due to impaired development of inferior vena cava (IVC). Some investigators also use the term preureteral vena cava considering the embryology. The ureter, in this condition, traverses behind the vena cava, partially circumscribes the medial edge of vena cava and pursues normal distal course. It occurs commonly on the right side, unless associated with partial or complete situs inversus [1, 2] or duplication of IVC [3], when it occurs on the left. Bilateral circumcaval ureter has also been reported. Majority of patients

present in the third or fourth decades with right flank pain, recurrent infections, hematuria and stone formation. Preoperative diagnosis is possible by intravenous urogram (IVU), spiral CT or magnetic resonance imaging (MRI). Management is surgical and involves restoring the normal course by transposition and reanastomosis of circumcaval ureter. The surgical management, in the last decade, has shifted from open surgical repair to minimally invasive surgeries.

Case Report

A 45-year-old male presented with flank pain and history of febrile urinary tract infection in the past. Physical examination was normal. Haematological and biochemical investigations were normal. Abdominal sonography showed moderate hydronephrosis of right kidney (Fig. 1). IVU (Fig. 2) revealed characteristic fish hook or shepherd's crook appearance of type 1 circumcaval ureter. Open surgical correction was planned, and the ureter was approached extraperitoneally with an anterior subcostal muscle-cutting incision. The ureter was traced both in its pre- and post-caval courses (Fig. 3) and was repositioned to its normal course by ureteropelvic anastomosis over a double J stent. The fibrotic retrocaval portion was excised. The retroperitoneal drain was removed after the drainage had dried to less than 25 cc. The postoperative recovery was uneventful. The stent was removed cystoscopically at 6 weeks, and patient is asymptomatic at 6 months of regular follow-up.

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Discussion and Review of Literature

Circumcaval ureter was first reported by Hochstetter in 1893. As the anomaly is due to anomaly of vena caval development

Fig. 1 Ultrasonography showing hydronephrosis of the right kidney

rather than the ureter, some use the term preureteral vena cava. Circumcaval ureter is an uncommon congenital anomaly with an autopsy incidence which is about 1 per 1000 [2]. It occurs three times more commonly in males and on right side unless, when associated with complete or partial situs inversus or duplication of IVC, when it is seen on left side [3]. Bilateral circumcaval ureters have been reported in the literature [4].

Embryology

At 4 to 7 weeks of development, IVC develops, by successive development and regression of subcardinal, supracardinal and

posterior cardinal veins. The right subcardinal vein forms the pre-renal IVC, the subcardinal-supracardinal anastomosis forms the renal segment and the right supracardinal vein forms the post-renal IVC. The persistence of posterior cardinal veins leads to the ureter getting trapped behind the developing vena cava, resulting in circumcaval ureter. Several variants in the anatomic and topographic presentations of circumcaval ureter have been described in the literature [2, 5]. Concurrent congenital nonvascular abnormalities have also been reported frequently in humans with retrocaval ureter, including glandular hypospadia, supernumerary lumbar vertebra, syndactylia and intestinal malrotation [6].

Clinical Diagnosis

Patients of circumcaval ureter present with flank pain, recurrent urinary tract infection and hematuria in the third or fourth decades. The flank pain is either due to hydronephrosis or to associated calculi. Hematuria is of varying degrees. Fillo et al.

Fig. 2 IVU showing hydronephrosis of the right kidney with 'seahorse' or 'shepherd's crook' appearance

Fig. 3 Operative photograph showing pre- and post-caval segments of circumcaval ureter

reported a case of gross hematuria, due to a carcinoma in a circumcaval ureter [7]. Hydronephrosis in circumcaval ureter is described as a consequence of ureteral compression by psoas muscle, spinal column and IVC [8, 9]. However, ureteral obstruction is not an inevitable complication [10].

Radiologic Diagnosis

Abdominal ultrasound can only be the screening radiologic investigation for those with circumcaval ureter which shows ipsilateral moderate hydronephrosis.

IVU is the most commonly used radiologic investigation to diagnose circumcaval ureter. Depending upon the IVU findings, Bateson and Atkinson classified circumcaval ureter into two types [9]. Type 1, also called ‘low loop’, is the most common type and seen in 90 % of patients and results in hydronephrosis in 50 % of these. The point of obstruction is placed some distance from lateral margin of IVC at the level of the third lumbar vertebra. The appearance has variously been described as seahorse sign, fish hook or shepherd’s crook appearance. Type 2 is less common, also called high loop with the site of obstruction being at a higher level, and results in sickle-like appearance. IVU has its limitation in that ureter distal to obstruction is often not visualised.

Pienkny et al. suggested that a 3D, volume-rendered CT scan in the excretory phase combined with diuretic renography could be a radiologic investigation of choice in these patients [11]. Spiral CT which can simultaneously outline the ureter and IVC, free of respiratory artefacts and can produce overlapping of images, may be another choice to confirm the diagnosis after ultrasound. The presence of vena cava lateral to the right pedicle of the third lumbar vertebra, which is seen in 94 % of patients with circumcaval ureter and only 6 % of normal individuals, is said to be the pathognomonic feature of circumcaval ureter on CT [12].

In 2002, Uthappa et al. reported the potential of MRI to diagnose circumcaval ureter with equal effectiveness of spiral CT but without the radiation risk [13]. The same has been confirmed by Muthusami et al. [14]. MRI can also be used in those patients with poor renal function as MR urogram, in pregnant women and in children.

Treatment

Circumcaval ureter is described as a congenital anomaly that requires surgical correction only in symptomatic patients [15]. Watchful waiting is advised for those patients without hydronephrosis, infection or stone formation, intervening only in those who develop symptoms or whose kidney function deteriorates.

Open surgical ureteroureterostomy is considered as a gold standard for surgical intervention. Division of dilated pelvis with transposition and reanastomosis, initially described by Harril in 1940 [16], is the most commonly practised intervention in symptomatic patients. In Harril’s method, ureteropelvic anastomosis is done after dividing pelvis just above pelviureteric junction, which will maintain the abundant vascular supply of pelvis as well as proximal ureter. The retrocaval portion of the ureter can either be excised or left alone. Xiaodong et al. suggested that if a 8-F catheter could not be passed, the segment should be excised [8]. On the contrary, Puigvert did the anastomosis leaving the retrocaval segment of the ureter unexcised [2, 17].

Goodwin et al. proposed in 1957 that the IVC be divided and reanastomosed after repositioning the ureter [18]. This is no longer practised, considering the surgical risks and better techniques available.

Xiaodong et al. suggested ‘vena cava supporter’ for those circumcaval ureters, accompanied by ureteral torsion and vena caval compression. A chemical or a metal tube covered by psoas muscle is placed between vena cava and ureter after correcting the anomaly [8].

After laparoscopic dismembered pyeloplasty was described by Schuessler et al. in 1993 [19], it was but natural that the technique be used for circumcaval ureter as well. It has the obvious advantage of minimal disfigurement and morbidity. Both transperitoneal and retroperitoneal approaches have been used and both have their advocates. The retroperitoneoscopic technique provides a direct access, without the division of lateral peritoneal reflection. Solomon et al. were the first to report purely retroperitoneoscopic technique in 1999 [20]. The total surgical time was prolonged mainly due to the time taken for intracorporeal ureteroureteral anastomosis. Mugiya et al. used an automatic suturing device to bring down the surgical time [21]. Zhang et al. in their series of 15 cases reported a surgical time of 120 min [22]. Hemal et al. reported a case of circumcaval ureter repaired by purely robotic technique and concluded that it did not offer any additional benefit over laparoscopic repair apart from better ergonomic ease for intracorporeal suturing [23]. Nayak et al. reported a series of five patients undergoing pure robotic correction of circumcaval ureter [24].

Conclusion

Circumcaval ureter is an uncommon congenital anomaly resulting in proximal hydronephrosis. In the last decade, there has been a paradigm shift in the diagnosis and surgical management from invasive to minimally invasive modalities with good surgical outcome.

Conflict of Interest The authors declare that they have no competing interests.

Consent Written consent was taken from the patient for publication of the case details and use of photographs.

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