Case Report

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Circumcaval Ureter with Vesico Ureteral Reflux: The First Association in Literature

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Abstract

A circumcaval ureter is a rare congenital anomaly in which the ureter passes behind, and is compressed by, the inferior vena cava. Its etiology is assumed to be abnormal embryologic development of the inferior vena cava as a result of atrophy failure of the right subcardinal vein in the lumbar portion. A circumcaval ureter is also termed a retrocaval ureter. The right supracardinal system fails to develop, whereas the right posterior cardinal vein persists. With one reported exception, the anomaly always occurs on the right side. Patients with this anomaly may develop partial right ureteral obstruction or recurrent urinary tract infections. Therapeutic options include surgical relocation of the ureter anterior to the cava.

A 14-year-old female patient came with complaints of fever, intermittent colic and dysuria 4 years ago. A right ureteric fourth-grade VUR and circumcaval ureter were established.

An anomaly in which both of these are together could not be found in literature. If after the VUR treatment he has progressive abdomen pain and advancing hydronephrosis, a circumcaval ureter as an additive anomaly must not be forgotten. For that reason, in a patient having a urinary system anomaly, a likely extra anomaly should be searched.

Key words: *Anomalies, circumcaval, hydronephrosis, vena cava inferior/abnormalities, vesico-ureteral reflux*

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Introduction

A circumcaval ureter is a rare congenital anomaly in which the ureter passes behind, and is compressed by, the inferior vena cava. Its etiology is assumed to be abnormal embryologic development of the inferior vena cava as a result of atrophy failure of the right subcardinal vein in the lumbar portion [1]. A circumcaval ureter is also termed a retrocaval ureter [2-3]. The right supracardinal system fails to develop, whereas the right posterior cardinal vein persists. With one reported exception [4], the anomaly always occurs on the right side. Patients with this anomaly may develop partial right ureteral obstruction or recurrent urinary tract infections. Therapeutic options include surgical relocation

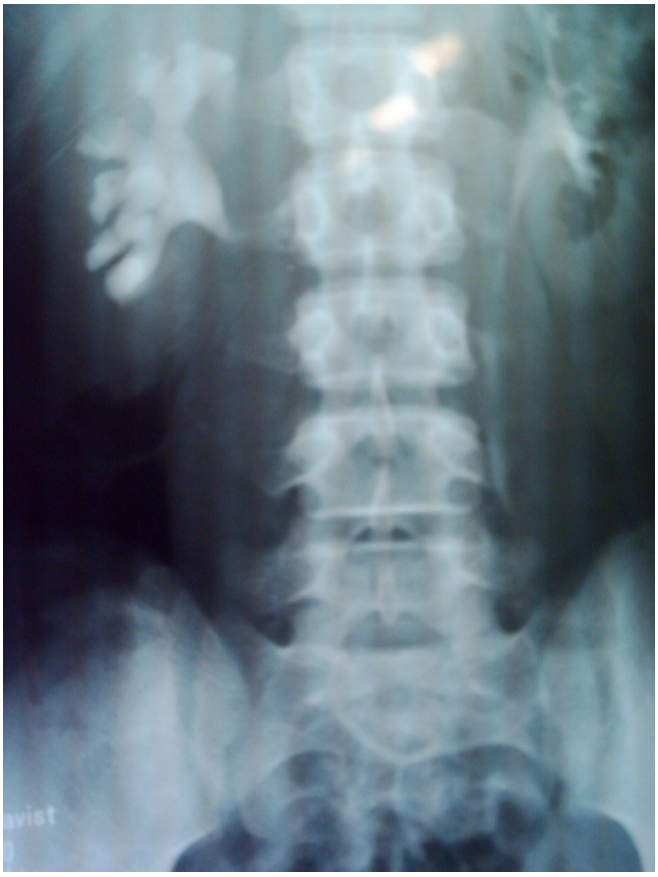

Figure 1. "J" or "Fish hook" appearance of pelvis and ureter.

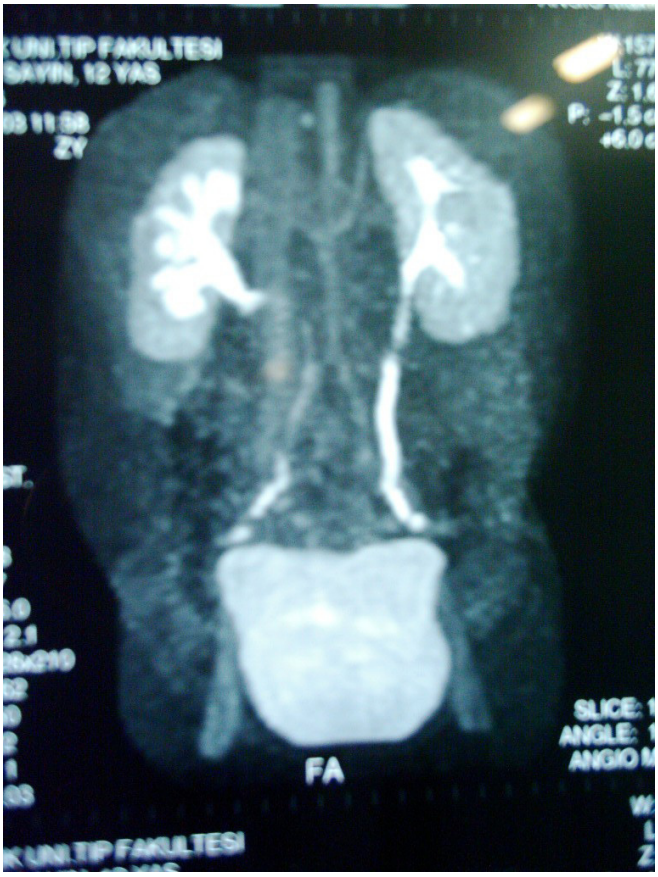

Figure 2. MR urography findings of circumcaval ureter.

of the ureter anterior to the cava [4].

Vesico-ureteral Reflux (VUR) is the abnormal flow of urine from the bladder into the upper urinary tract. The prevalence of vesico-ureteral reflux in normal children has been estimated to be 0.4% to 1.8%. Based on epidemiological data, it can be estimated that 2.2% of girls and 0.6% of boys may be found to have reflux as a result of the investigation of urinary tract infections [5]. In the majority of cases, it occurs as a result of a primary maturation abnormality of the vesico-ureteral junction or a short distal ureteric submucosal tunnel in the bladder that alters the function of the valve mechanism [6]. Familial clustering of VUR implies that genetic factors have an important role in the pathogenesis of reflux [7].

Case Report

A 14-year-old female patient came with complaints of fever, intermittent colic and dysuria 4 years ago. She was hospitalized after a diagnosis of pyelonephritis and was treated. In her investigations carried out after treatment, a right ureteric fourth-grade VUR was established in voiding cystourethrography and the VUR was repaired surgically. During the observations of the patient, she was examined again because of abdomen pain. Ultrasonographic examination revealed right hydroureter and hydronephrosis. We suspected a circumcaval ureter on intravenous urography (IVU) findings of an inverted "J" or "Fish hook" appearance of the pelvis and ureter (Figure 1) as well as MR urography findings of a circumcaval ureter (Figure 2). The patient had a Type l form of a retrocaval ureter. During operation, the renal parenchymal surface was normal. The proximal segment of the ureter was varicose. The retrocaval portion showed severe adhesions. It did not admit a 4F ureteric catheter. The distal ureter was normal. The ureter was severed from the renal pelvis (Figure 3) and a dismembered pyeloplasty was performed with the distal segment leaving the retrocaval segment in-situ. Moreover, patient pain decreased in the postoperative period and hydronephrosis improved (Figure 4).

Discussion

The circumcaval ureter is a rare congenital anomaly caused by an error in the embryogenic development of the inferior vena cava (IVC). The concomitant abnormalities were mainly related to the two implicated organs, vena cava and kidney, and generally with the two

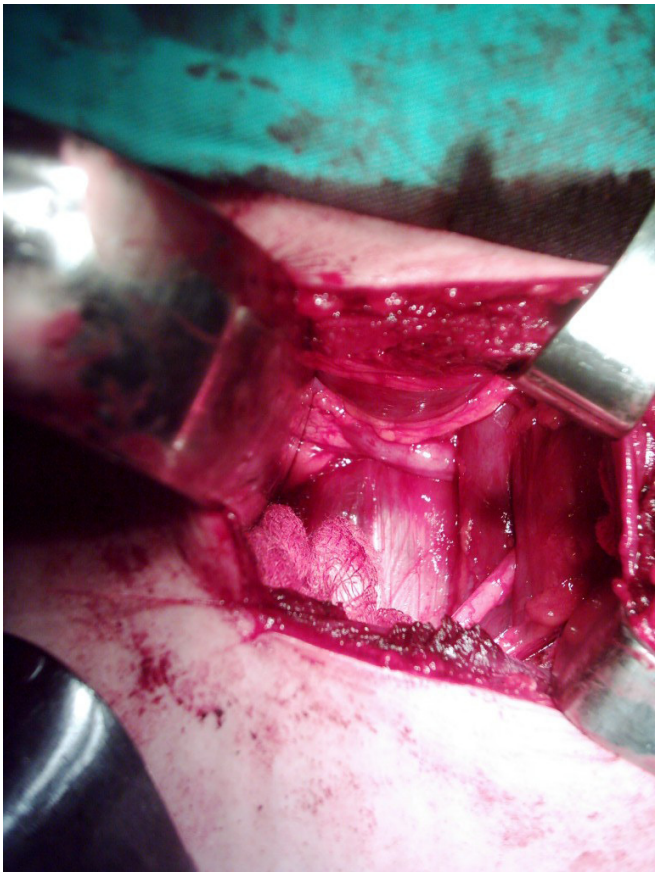

Figure 3. The ureter was severed from the renal pelvis.

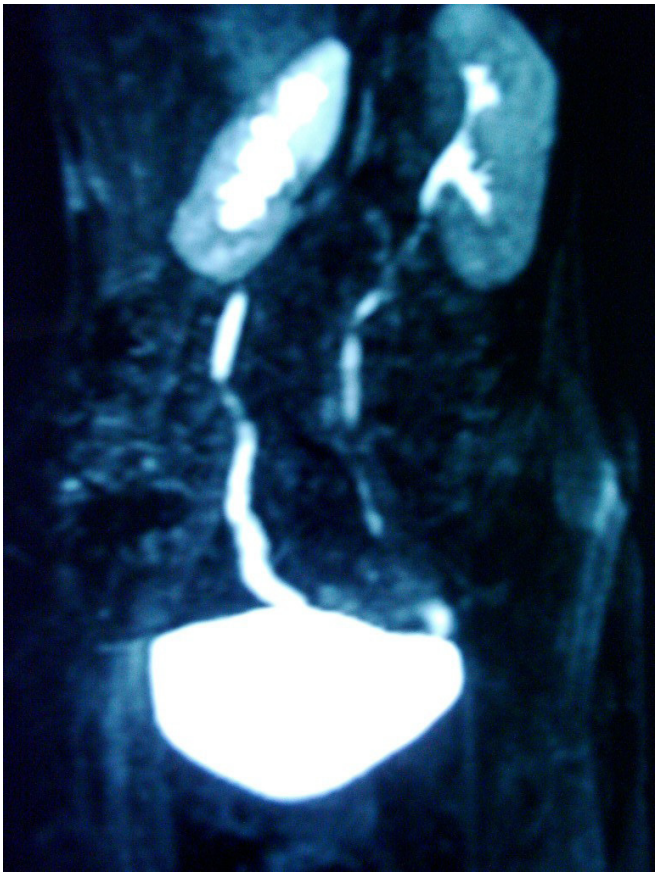

Figure 4. Postoperative period and hydronephrosis improved.

implicated systems, i.e. cardiovascular and genitourinary. Overall, it appears that about 20% of the patients with a retrocaval ureter present concomitant congenital abnormalities, some of which would be useful to be known [8]. It is usually associated with upper urinary tract hydronephrosis, and patients present in their third to fourth decade of life with right flank pain and discomfort.

Patients may also complain of urinary tract infections, hematuria, or fever.

Circumcaval ureters have been classified into two clinical types across the literature, in accordance with the common radiological appearance [2, 9]. Type 1 (also named "low loop") has been reported as the more common form; it is characterized by the so-called "typical S" or "fishhook" deformity of the ureter to the level of the obstruction, with the point of obstruction placed some distance from the lateral margin of the IVC at the level of the third lumbar vertebra. In the type 2 variant (also called "high loop"), the ureter has a "sickle-shaped" curve, with the point of obstruction at the lateral margin of the IVC. This second variant is rare, and represents around 10% of the known cases [10].

Type 1 is usually associated with moderate to severe hydronephrosis in 50% of the patients and type 2 with mild or no hydronephrosis [9]. Indeed, in the latter form, the upper ureter is nonkinked but passes behind the IVC at a higher level; the renal pelvis and upper ureter lie almost horizontal before they encircle the IVC with a smooth curve. The ureter is compressed against the perivertebral tissue [9].

Most patients complain of right flank pain with recurrent urinary tract infections due to ureteral obstruction, or an acute right pyelonephritis as the next most common cause of presentation. Symptoms may be attributable to calculi, and hematuria is frequently present [3, 9].

Various techniques for the management of a circumcaval ureter have been reported. In patients with minimal caliectasis and no subjective symptoms, surgical correction is not mandatory, but observation should be maintained. Therefore, a circumcaval ureter has been defined as a rare congenital anomaly that requires surgical correction in the symptomatic patients. Conservative treatment and periodical examination should be given to those patients without hydronephrosis, in-

fection, and stone formation. A periodic examination has also been suggested for patients with mild hydronephrosis [11-12].

The choice of an open surgery approach primarily depended on the severity of the hydronephrosis, impairment of kidney function, and the type of anomaly [13]. Most of the authors suggested an ureteropelvis anastomosis by which a section is made at the level of the pelvis just above the ureteropelvic junction. The performance of laparoscopic dismembered pyeloplasty in the setting of a retrocaval ureter leads to added challenges, including extensive caval dissection, both lateral and medial to the vena cava. Despite these challenges, this procedure can be optimally performed via a minimally invasive approach. We present below a case of a retrocaval ureter treated with laparoscopic dismembered pyeloplasty [14].

The prognosis is good and complications are uncommon after surgery in childhood; however, recovery is slower in adults due to compliance of the pelvis from long-term higher intrapelvic

pressure. As a circumcaval ureter is seldom symptomatic in children, it is often misdiagnosed

for years. It is generally accepted that the low incidence in children is a reflection of the gradual increase of the associated hydronephrosis [15].

In our patient, grade IV VUR was established as well as a circumcaval ureter. VUR is a urinary system anomaly. An anomaly in which both of these are together could not be found in literature. If after the VUR treatment a patient has progressive abdomen pain and advancing hydronephrosis, a circumcaval ureter as an additive anomaly must not be forgotten. For that reason, in a patient having a urinary system anomaly, a likely extra anomaly should be searched.

Conflict of interest statement

The authors declare no competing interest. No financial support was received for this paper.

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