Unusual presentation of retrocaval ureter with recurrent vomiting: A case report

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ABSTRACT

Retrocaval ureter is a rare congenital anomaly in which the ureter takes an abnormal course from behind the inferior vena cava (IVC). A 26-year-old male was referred to Urology outpatient clinic due to recent finding of right sided hydronephrosis on renal ultrasound and recurrent episodes of vomiting lasting for 2–3 days for the past 4–5 years. On conducting further imaging studies, a diagnosis of retrocaval ureter was made. Open surgical reconstruction was successfully carried out, resulting in complete resolution of the patient’s symptoms. In this case report, we highlight an unusual presentation of this relatively uncommon congenital anomaly.

Introduction

Retrocaval ureter is a rare congenital anomaly most commonly affecting the right ureter due to aberrant development of the IVC. It has a worldwide incidence of 0.06%–0.17% with a little over 200 cases reported since 1893. It usually presents with abdominal pain, flank pain or, hematuria. Our case depicts an unusual presentation of recurrent episodes of vomiting caused by this relatively rare developmental abnormality.

Case presentation

A 26-year-old male was referred to Urology outpatient clinic due to recent finding of hydronephrosis of right kidney on ultrasound. The patient had a history of recurrent episodes of vomiting occurring monthly for the past 4–5 years with each episode lasting for 2–3 days. The episodes occurred almost every month and the patient was symptom free during the intermittent period. Lab investigations and imaging studies including abdominal ultrasound and upper GI endoscopy conducted in the past were reported to be normal and the patient was prescribed antiemetics for cyclical vomiting syndrome/anxiety neurosis. Subsequently, CT urography revealed retrocaval ureter involving right proximal ureter (Fig. 1). A retrograde pyelogram was done for better delineation and showed characteristic fishhook shaped or S-shaped deformity (Fig. 2). Open surgical reconstruction through right subcostal approach was planned. Dilated renal pelvis was transected. Ureter was transposed anterolateral to the inferior vena cava. Redundant segment of dilated proximal ureter and retrocaval segment was excised. Ureter was spatulated and anastomosed with renal pelvis over a DJ stent. The patient was followed regularly for one year during which he consistently reported complete resolution of his symptoms.

Discussion

Retrocaval ureter is a rare congenital malformation with an incidence of 0.06–0.17%. Since its discovery by Hochstetter in 1893, only about 200 cases have been reported worldwide. It is three times more common in men as compared to women and patients usually present in 3rd – 4th decade of life, as seen in our case.

Retrocaval ureter in reality is a congenital malformation of the inferior vena cava. The infrarenal IVC instead of arising normally from the posteriorly located supracardinal vein, arises from anteriorly located subcardinal vein. This results in the entrapment of proximal part of ureter between the IVC and Psoas muscle leading to it’s kinking and formation of an adynamic segment which causes hydronephrosis. Often, patients are either asymptomatic or present with abdominal pain, flank pain and hematuria. Complications such as urinary infection, stone formation and renal dysfunction can also arise due to stasis of urine. In contrast to the classical presentations of retrocaval ureter, the episodic vomiting experienced by our patient can probably be explained by reno-gastric reflex. Intermittent rapid distension of renal pelvis due to ureteric compression can increase the pressure of pyloric sphincter and lead to vomiting. This unusual presentation led to a delay in diagnosis. Moreover, the patient was erroneously thought to have either abdominal

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migraines, cyclical vomiting syndrome, food allergies or, emotional disturbances due to normal lab findings initially.

Diagnosis of retrocaval ureter is made by either ureterography (intravenous or retrograde) or CT Urography. Based on the level of obstruction on imaging, retrocaval ureter is of two types. In type 1 retrocaval ureter, which is the more common type, the ureter crosses behind the IVC at the level of L3/L4 vertebra and there is marked hydronephrosis. This leads to classic appearance of ureter as fishhook shaped or S-shaped on ureterogram as seen in our case. Compared to type 1, the ureter crosses the IVC higher at the level of renal pelvis in type 2. This leads to minimal hydronephrosis and the ureter appears sickle shaped on ureterogram.⁷

Surgery for retrocaval ureter is indicated when either there is significant functional obstruction, pain or renal function deterioration. It can be done either by open surgical reconstruction or by laparoscopic repair.⁷ In our patient, surgery led to complete resolution of his symptoms as noted on subsequent follow up visits.

Conclusion

With this case report, we aim to illustrate an atypical presentation of this rare congenital malformation. Retrocaval ureter should be considered as a possible cause of unexplained recurrent vomiting episodes especially in a young male adult.

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Fig. 1. CT urography showing retrocaval ureter.

Fig. 2. Retrograde pyelography showing fishhook shaped or S-shaped deformity.