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Case report

Retrocaval ureter with vesicoureteric reflux, a very rare entity

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Right subcardinal vein;
Ureteroureterostomy

Abstract
Introduction: A circumcaval ureter is a rare congenital anomaly in which the ureter passes behind inferior vena cava. VUR is rarely found in association with retrocaval ureter. Diagnosis and management are difficult. To our knowledge, we report a second case of retrocaval ureter with ipsilateral VUR.
Observation: A 9-year-old child was admitted with complaints of right renal pain. After workup a diagnosis of retrocaval ureter with VUR was made. He underwent ureteroureterostomy with excision of retrocaval segment and is being managed conservatively for low grade VUR.
Conclusion: In a child presenting with retrocaval ureter we should look for associated VUR. The problem is similar to pelviureteric junction obstruction and ipsilateral reflux. The management is to deal with obstruction first, followed by extravesical ureteric reimplanation or subureteric teflon injection for reflux.

Introduction

Retrocaval ureter is a rare congenital anomaly in which the ureter passes behind the inferior vena cava. It results due to failure of the lumbar segment of right subcardinal vein to get atrophied [1]. The incidence of retrocaval ureter is 1 in 1500 cadavers; male to female ratio is 3 or 4:1 [2–4]. A total of 20% of cases of retrocaval ureter present with concomitant anomalies, mainly from cardiovascular system and genitourinary tract. Associated anomalies are right double inferior cava, ipsilateral ureterocele, glandular hypospadias, supernumerary lumbar vertebrae, syndactylia, partial situs inversus,

Abbreviations: VUR, vesicoureteric reflux; DJ, double J; IVU, intravenous urogram; USG, ultrasonography; MCU, micturating cystourethrogram; MRU, magnetic resonance urography.

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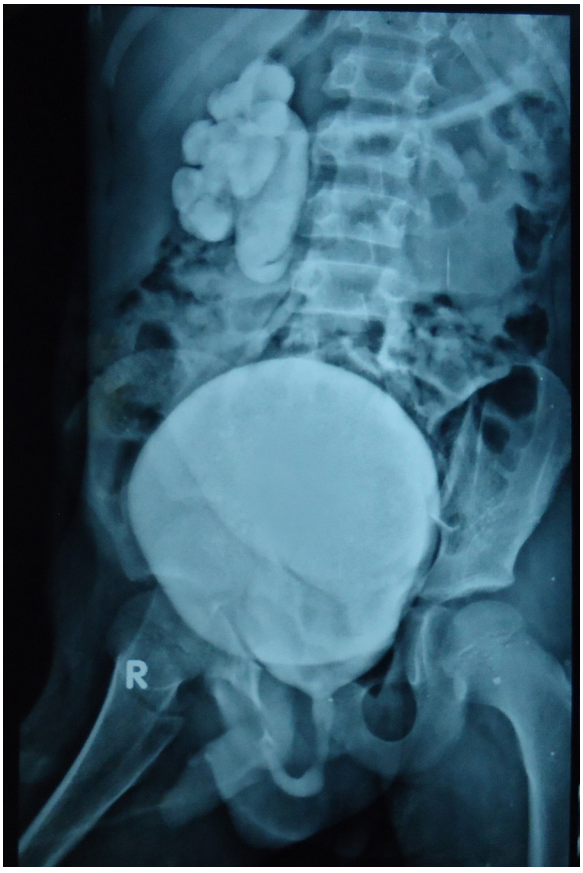

Figure 1 MCU having grade 4 right side vesicoureteric reflux.

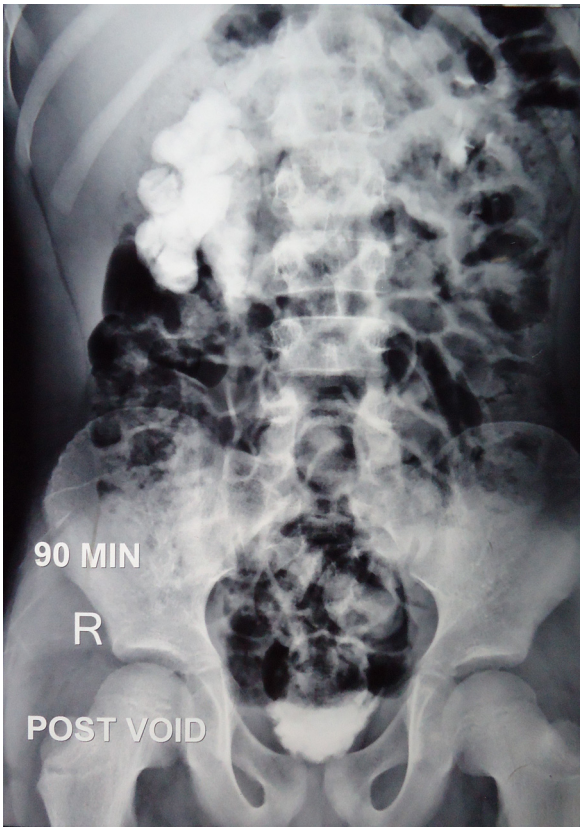

Figure 2 IVU showing right pelviureteric junction obstruction.

etc. [5]. The prevalence of vesicoureteric reflux (VUR) in boys is 0.6% [6]. We present a case of retrocaval ureter with VUR on the same side. On searching the literature we could find only one case report of such a combination. So, rarity of the case and its management problems warrant this publication.

Case report

A 9-year-old boy was admitted with complaints of right renal pain for one month with no urinary complaints or fever in May 2015. Physical examination was unremarkable. CBC, blood urea and serum creatinine were normal. Urine culture was sterile. Ultrasonography (USG) of abdomen showed right gross hydroureteronephrosis. Micturating cystourethrogram (MCU) revealed grade 4 VUR (Fig. 1 – MCU showing grade 4 right side vesicoureteric reflux). Intravenous urogram (IVU) showed diagnostic dilemma between pelviureteric junction obstruction and retrocaval ureter (Fig. 2 – IVU suggestive of pelviureteric junction obstruction). For confirmation of the diagnosis magnetic resonance urography (MRU) (T2 weighted) was done confirming type 1 retrocaval ureter (Fig. 3 – vertical thick arrow indicates IVC while horizontal thin arrow indicates ureter). The patient underwent open ureteroureterostomy with excision of retrocaval segment with double J (DJ) stenting (Fig. 4 – vertical arrow indicates IVC while horizontal arrow shows retrocaval ureter segment). DJ stent was removed 4 weeks later. At 6 weeks MCU showed right side low grade VUR and residual dilatation of pelvicalyceal system and upper ureter (Fig. 5 – right side reflux with minimally dilated lower ureter and residual hydroureteronephrosis).

At 6 months follow-up he is asymptomatic. Urine culture was sterile at 1, 3, 6 and 9 months follow-up. On USG hydronephrosis has not increased.

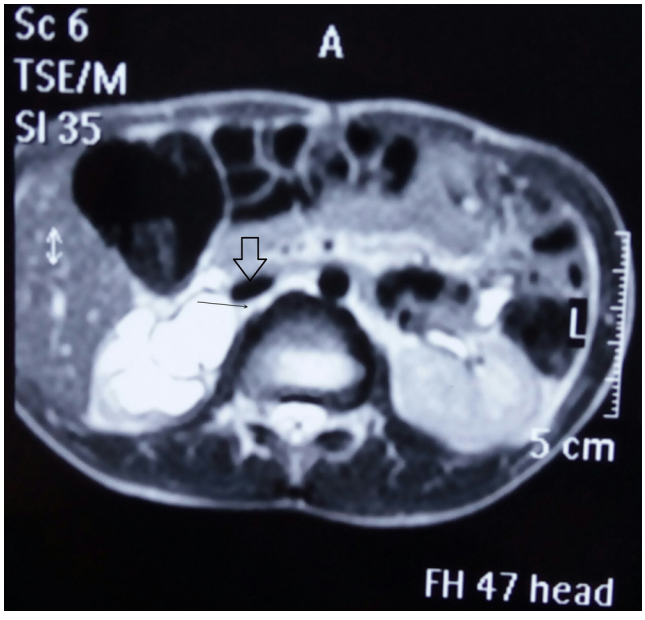

Figure 3 MRU depicting right retrocaval ureter (vertical thick arrow indicates IVC while horizontal thin arrow indicates ureter).

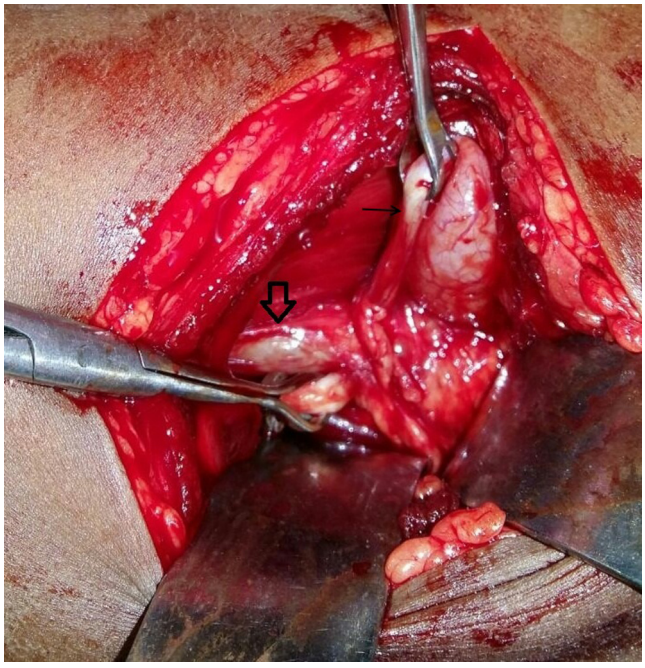

Figure 4 Retrocaval ureter was excised surgically by open ureteroureterostomy vertical arrow indicates IVC while horizontal arrow showing retrocaval ureter segment.

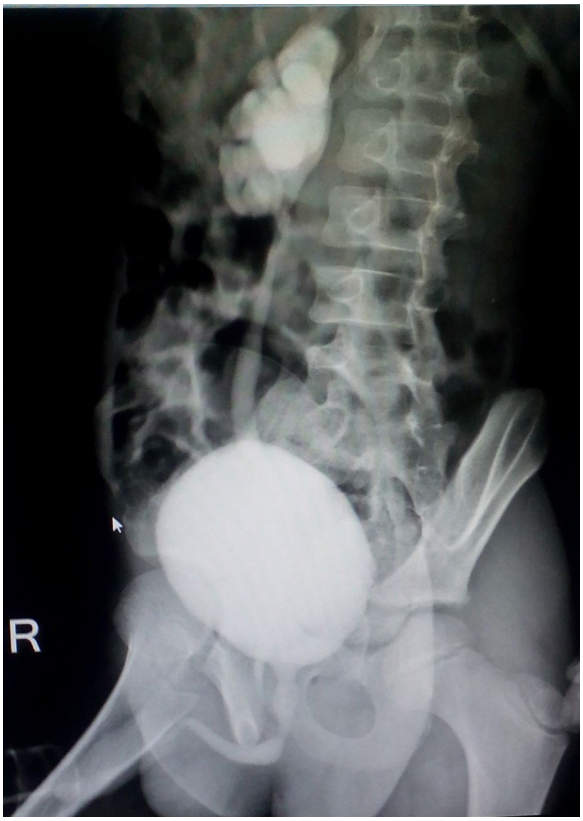

Figure 5 Postoperative MCU shows right side VUR with minimally dilated lower ureter and residual hydroureteronephrosis.

Discussion

Retrocaval ureter is also called circumcaval ureter resulting from persistence of posterior cardinal vein or failure of regression of right subcardival vein during embryologic development. Retrocaval ureter is of 2 types. Type 1 is fish hook or s-shaped. Ureter passes behind the inferior vena cava and lateral to lumbar vertebrae to cross anteriorly and assumes a normal distal course. Type 2 is sickle-shaped and less angulated than type 1. Ureteral obstruction is extrinsic; however, in some cases intrinsic ureteral fibrosis has been documented. Most of the cases occur on right side except in situs inversus/duplication of inferior vena cava [7,8]. Since it is associated with multiple congenital anomalies, one should also look for them. All affected individuals are not symptomatic. Patients usually present with right ureteral obstruction, recurrent urinary tract infection or hypertension in third or fourth decade of life.

IVU findings may mimic ureteropelvic junction obstruction. MR urogram or CT urogram is diagnostic. Diuretic renogram may be needed to document obstruction and split renal function. Patients presenting with symptoms and documented obstruction are candidates for surgical intervention. Surgery includes excision of retrocaval ureter segment and anterior relocation of ureter by open or laparoscopic approach. The retrocaval segment should always be excised as it may be intrinsically abnormal [9].

VUR is retrograde flow of urine from bladder into the ureter or pelvicalyceal system. Primary VUR is a congenital malformation resulting from short intramural ureteric tunnel. MCU is the diagnostic study. Five grades of VUR have been described based on MCU images. Lower grades tend to resolve spontaneously. High grades of reflux may be associated with renal scarring.

The literature cited only one case of retrocaval ureter with ipsilateral primary VUR in a 14-year-old girl [10]. Herein we present a 9-year-old boy who presented with right renal pain. IVU suggested right ureteropelvic junction obstruction. MCU showed grade 4 ipsilateral VUR. MR urogram confirmed type 1 right retrocaval ureter on T-2 weighted images.

The association of retrocaval ureter and VUR is challenging for both diagnosis and management. This situation is akin to ureteropelvic junction obstruction and VUR on same side. Both the problems cannot be managed in one sitting as we cannot dismember ureter at 2 places. Ciftci et al. treated VUR first as the girl presented with recurrent UTI. However, he did not provide the details of the surgery [10]. Later he did pyeloureterostomy for upper urinary tract symptoms. In the present case, we decided to treat retrocaval ureter first as he came with upper ureteral obstruction. Post-operative MCU showed minimal lower ureteral dilatation and residual hydronephrosis. His urine culture was sterile and there was no history of documented urinary tract infections. Therefore, we are following up with options of extravesical ureteric reimplanation or subureteric teflon injection in case he presents with recurrent urinary tract infection.

In these types of situations we should treat the obstructive lesion first, followed by treatment of reflux if needed, without severing the ureter.

Conclusion

We have probably reported a second case of retrocaval ureter with ipsilateral VUR. Though rare, one should look for this association. Diagnosis and treatment is challenging. Obstructing lesion should be managed first.

Authors' contributions

All authors are working in Department of Urology, SP Medical College, Bikaner, and contributed for case management and preparation of the manuscript.

Ethical committee approval

Not required.

Patient's consent

We took written consent from patient's parents.

Conflict of interests

None declared.

Source of funding

None.

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