Congenital venous anomalies associated with retrocaval ureter: evaluation using computed tomography

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[Received: 1 February 2022; Accepted: 7 March 2022; Early publication date: 5 April 2022]

Background: Retrocaval ureter is a rare congenital anomaly resulting from anomalous development of inferior vena cava (IVC) and not from anomalies of the ureter. The anomaly always occurs on the right side due to regression of right supracardinal vein and persistence of right posterior cardinal vein. Retrocaval ureter tends to be associated with various vena cava anomalies because of the embryogenesis. We aimed to identify the prevalence of associated congenital venous anomalies (CVA) resulting from cardinal vein development in adults with retrocaval ureter using computed tomography (CT) images.

Materials and methods: The study included 22 adults with retrocaval ureter. We evaluated CT findings and determined the incidence of associated CVA using thin slice data sets from CT scanner with 64 or more detectors. We compared the prevalence of CVA in the retrocaval ureter group (mean age: 57 ± 19 years) and in the control group of 6189 adults with normal ureter (mean age: 66 ± 14 years).

Results: In the retrocaval ureter group, 4 (18.2%) adults had CVA including double IVC, right double IVC, preisthmic IVC with horseshoe kidney, and preaortic iliac confluence. One of 2 adults with preaortic iliac confluence had right double right IVC. In the control group, 49 (0.79%) adults had CVA including 37 double IVC, 11 left IVC, and 1 IVC interruption azygos continuation. Fifteen horseshow kidneys were found. The prevalence of associated CVA in the retrocaval ureter group was higher than that in the control group (p < 0.001).

Conclusions: Retrocaval ureter is frequently associated with CVA. Various CVA with retrocaval ureter could happen because of abnormal development of not only the right posterior or supra cardinal vein but also other cardinal veins. (Folia Morphol 2023; 82, 2: 300–306)

Key words: retrocaval ureter, vena cava, congenital venous anomaly
INTRODUCTION

Retrocaval ureter, also known as circumcaval ureter, or preureteral vena cava, is a congenital condition characterised by the persistence of the posterior cardinal vein on the right, which causes the proximal ureter to deviate medially, behind the inferior vena cava (IVC), before resuming its natural course anteriorly and laterally [1, 9, 13, 14, 17]. Many authors prefer using the term “preureteral vena cava”, as the cause of this variant is IVC developmental abnormality, and not a ureteral one [9, 13–16, 21]. The prevalence of retrocaval ureter was reported as 0.06–0.27% and the overall prevalence was 0.13% [2, 14]. Males are affected by retrocaval ureter about 3 times more often than females [2, 4]. Bateson and Atkinson distinguished two types of retrocaval ureters: type 1 of low loop (S or ‘fish-hook’ deformity), in which the ureter crosses behind the IVC at the level of the L3 vertebra, and type 2 of high loop (sickle-shaped deformity), in which the renal pelvis and the upper ureter lie horizontally [2]. The low loop type is more common than the high loop type and has a moderate or severe hydronephrosis [2]. From a clinical standpoint, many cases of retrocaval ureter are asymptomatic, and only detected incidentally using imaging techniques [9, 13, 16]. Computed tomography (CT) elegantly depicts the abnormal course of the ureter [13]. When present, the symptoms are usually abdominal pain and haematuria due to ureteral obstruction or urinary infection [1, 2, 13, 16, 26]. Retrocaval ureter has been associated with other local or general congenital abnormalities including horseshoe kidney, right double IVC, contralateral kidney agenesis, preaortic venous confluence [1–3, 7, 9–16, 20, 21, 23, 27, 30]. The associations are related to the development of cardinal veins. A review of the literature by Perimenis et al. [20] revealed that 21% of the cases of retrocaval ureter present with concomitant abnormalities mainly from the cardiovascular system and the genitourinary tract. There are many case reports about congenital association; however, the prevalence of the associated congenital venous anomalies (CVA) and clinical features were not evaluated in case series. The purpose of this article was to identify the prevalence of the associated CVA in patients with retrocaval ureter using CT data, and to emphasize its clinical importance.

MATERIALS AND METHODS

The institutional ethics committee approved this retrospective study and granted a waiver for the requirement of informed consent.

Our study included 22 adults with retrocaval ureter (18 males and 4 females, mean age: 57 ± 19 years old) and 6189 adults with a normal urinary system (3382 males and 2807 females, mean age: 66 ± 14 years old) in the control group. The control group was selected using the inclusion and exclusion criteria depicted in Figure 1. Cases with retrocaval ureter were searched using the key words “retrocaval ureter”, “circumcaval ureter”, “postcaval ureter” and “preureteral vena cava” among the abdominal CT reports of adults between January 2008 and September 2020 in our university hospital and affiliated hospitals. We excluded cases examined using non-contrast CT. Twenty-two adults with retrocaval ureter underwent contrast enhanced chest-to-abdominal CT between January 2008 and September 2020 in our university hospital and affiliated hospitals. CVA resulting from cardinal vein development and congenital renal anomaly (CRA) were evaluated using CT images (0.5–3 mm) on a picture archiving and communication system (PACS) workstation (SDS viewer, NOBORI Ltd. Tokyo, Japan). For retrocaval ureter group, acquired renal diseases were assessed using CT and medical records. We evaluated side and shape of the retrocaval ureter. We used classification of retrocaval ureter on the shape by Bateson and Atkinson [2].

Computed tomography was performed using a 64- to 128-slice scanner (SOMATOM Force, SOMATOM Definition Flash, SOMATOM Definition Edge, Siemens...
AG, Munich, Germany) at a slice thickness of 0.5 mm. The other parameters were as follows: tube voltage, 60–120 kVp; tube current, auto mA; and rotation time: 0.5 s. Contrast-enhanced CT examinations were performed by injecting 2 mL/kg of non-ionic contrast material at a rate of 2 mL/s with scanning delay of 120 s. CT urography was performed with more than 300 s of scanning delay.

Two radiologists with more than 20 years’ experience each in CT image interpretation reviewed 1 mm reconstructed axial CT images on a PACS workstation. If needed, additional multiplanar reformations, maximum injection projection and CT urography were used for the evaluations. The radiologists resolved any disagreement through discussion to reach a consensus.

We compared the prevalence of associated CVA and CRA in the retrocaval ureter and control groups using the Mann-Whitney U test. Demographic data in adults with CVA was compared between two groups by chi-square test. Statistical analysis was performed using SPSS version 23 software. P values < 0.05 were considered statistically significant.

RESULTS

In the retrocaval ureter group, 4 adults with CVA and CRA were found: 1 right double IVC (Fig. 2), 1 double IVC, 1 preisthmic IVC with horseshoe kidney (Fig. 3), and 2 cases of preaortic iliac confluence (Fig. 4). An adult with right double IVC had preaortic iliac confluence. In the retrocaval ureter group, the prevalence of CVA and CRA per person was 18.2% (4/22) and 4.5% (1/22).

The adults with CVA and CRA in the control group were 34 males (CVA: 24, CRA: 10) and 30 females (CVA: 25, CRA: 6), and the mean age was 59 ± 21 years old. The control group included 49 adults with CVA. All cases were IVC anomalies: 11 left IVC (8 males, 3 females), 37 double IVC (16 males, 21 females), 1 IVC interruption azygos continuation (1 female). Sixteen adults with CRA were found: 15 horseshoe kidneys (10 males, 5 females) and 1 right renal absence (1 female). One adult with double IVC was associated congenital right renal absence and bicornuate uterus. The prevalence of CVA per person was 0.79% (49/6189), including left IVC (0.18%), double IVC (0.58%), and IVC interruption azygos continuation (0.02%). The prevalence of CRA per person was 0.26% (16/6189).

The prevalence of CVA and CRA per person in the retrocaval ureter group was higher than that in the control group (p < 0.001).

Male ratio associated with CVA in the retrocaval ureter group was higher than that in the control group (p = 0.009). There was no significant differ-
Five adults with retrocaval ureter were evaluated by CT urography. The clinical and CT findings are shown in Table 1. All retrocaval ureters were on the right side with 14 low loop types and 8 high loop types. Fourteen adults including 4 with CVA in the retrocaval ureter group had asymptomatic and retrocaval ureter associated CVA were found incidentally. Seven adults with hydroureter were found and the patients’ symptoms were flank pain and haematuria. Eight adults had acquired renal diseases. Three of them treated with surgery, but retroperitoneal laparoscopic approach was not performed. One adult experienced recurrent hydronephrosis with ureter stone after surgery.

**DISCUSSION**

Retrocaval ureter tends to be associated with IVC anomaly due to the embryological malformation of the cardinal vein [9, 13–16, 21, 27]. Ours is the largest study of associated CVA and CRA in adults with retrocaval ureter using CT data. Our results show that the prevalence of associated CVA was 18.2%. The prevalence of congenital IVC anomalies in the general population was reported as 0.2–3.0% [3–5]. In our study the prevalence was 0.79% and the result was similar to that on previous study. The prevalence of associated CVA and CRA was significantly more frequent in adults with retrocaval ureter than those with a normal ureter. The review of literature by Perimenis et al. [20] revealed that 21% of the cases...
of retrocaval ureter present with congenital abnormalities mainly from the cardiovascular system and the genitourinary tract. Present result was similar to that of the review article. However, they did not assess associated congenital cardiovascular diseases in detail [20]. They reported 9 horseshoe kidneys in 352 cases with retrocaval ureter with prevalence of 2.6%, and the prevalence of CRA including renal agenesis was reported as 5.1% [20]. The prevalence of CRA was 4.5% in our study; however, no renal agenesis was found, likely because our sample was small.

Inferior vena cava anomalies in retrocaval ureter patients were left IVC, double IVC, double right IVC and preisthmic IVC with horseshoe kidney [4–7, 10–12, 15, 18, 20–24, 30]. Retrocaval ureter is usually found on the right [1, 2, 9, 13, 14, 16]. All retrocaval ureters found in our study were on the right. Left retrocaval ureter is associated with situs inversus, or duplicated or single left IVC [4, 5, 7, 20, 21, 28, 30]. In double IVC, the retrocaval ureter is present on the right or left side [4, 5, 7]. The double IVC case in our study had the right retrocaval ureter. Right double IVC is an extremely rare condition, although there are some case reports describing it [5, 6, 11, 23, 24]. Right double IVC is sometimes associated with retrocaval ureter as we observed [6, 11, 23]. In the partial right double IVC, the anomalous ureter crosses thorough IVC split [6, 18].

Preisthmic IVC with retrocaval ureter is the specific finding and associated with horseshoe kidney [10, 12, 20, 29]. Because embryogenesis of the renal parenchyma and its venous drainage in the IVC occur simultaneously, it is plausible that horseshoe kidney and IVC anomalies are consequences of a shared disturbed signal that occurs as these retroperitoneal structures’ development [10, 12]. Impairment of renal ascent and rotation may affect the usual venous development [12].

Combined anomaly of retrocaval ureter and preaortic iliac confluence have been rarely report-

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<td>Urolithiasis</td>
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Cr — creatinine; CT — computed tomography; RCU — retrocaval ureter; ARD — acquired renal disease; CRA — congenital renal anomaly; CVA — congenital venous anomaly; HSK — horseshoe kidney; IVC — inferior vena cava; PVC — preisthmic IVC; DRIVC — double right IVC; DIVC — double IVC; PIC — preaortic iliac confluence; M — male; F — female; N — negative; P — positive
ed [6, 9, 24]. In normal development, the posterior aspect forms the iliac venous confluence, but persistence of the subcardinal vein results in preaortic iliac venous confluence [24]. Shin et al. [23] reported a case of right double IVC with combined retrocaval ureter anomalies and preaortic iliac confluence, as we observed.

Congenital IVR anomalies with normal ureter are more than half of left IVC on normal situs and most of them are double IVC, left IVC and IVC interruption azygos continuation [1, 13, 14]. Almost all retrocaval ureter presents on the right side because of the abnormal development of the right posterior cardinal vein. Right retrocaval ureters with double IVC and preaortic iliac confluence such as those in this study have been reported [7, 24]. Various CVA with retrocaval ureter could happen because of abnormal development of not only the right posterior cardinal vein but also other cardinal veins. Variations of associated congenital IVC anomalies were different in patients with normal or retrocaval ureter.

Associated congenital anomalies of the urinary system excluding horseshoe kidney are contralateral renal agenesis, ectopic or malrotated opposite kidney, hypospadias and absence of vas deferens [3, 9]. In our study, we only found a horseshoe kidney.

Retrocaval ureter occurs more often in men, which was confirmed by our results [9]. Retrocaval ureter is usually asymptomatic [9, 20]. In our study, 16 cases were incidentally found in CT examinations (76.2%). When present, symptoms most often begin at ages 30–40 [2, 26]. The mean age of our 5 symptomatic patients was 50 years old. Even though this condition is usually diagnosed in adults, there has been an increased number of case reports in recent years showing symptomatic cases in children [9]. Some patients had abdominal pain and haematuria due to hydronephrosis or urinary infection [1, 9, 13, 14, 16]. Associated urolithiasis and ureteral cancer were reported [8, 19, 25]. Surgical management is needed when the patient is symptomatic with documented subrenal functional obstruction [1, 8, 13, 16, 19, 25]. Surgical treatment should be performed as soon as possible when severe hydronephrosis is present and the upper ureter exhibits obvious dilation that affects the function of the kidney [9]. Patients with recurrent infection, secondary stones, and bleeding require urgent surgical treatment [9]. Surgical treatment involves transection and relocation of the ureter anterior to IVC [9, 16]. Laparoscopic reconstruction technique is effective and minimally invasive [16, 19, 20].

In our study, 3 patients (2 patients with renal stone and 1 patient with ureteral cancer) were treated with surgery. Two patients with mild hydronephrosis were observed. The main causes for hydronephrosis are stenosis or adhesion of the retrocaval segment and torsion [2]. All our retrocaval ureters associated with CVA were low type and no hydronephrosis was seen. Degree of retrocaval ureter compression in preisthmic IVC is unclear because the anomaly is very rare. Retrocaval ureter compression in the partial right double IVC might be severe because ureter crosses through its narrow slit. Careful observation is necessary for patients with low loop retrocaval ureter because there is marked hydronephrosis in up to 50% of the patients [2]. Caval dilatation due to aging might increase the risk of retrocaval ureter compression. Furthermore, complex anatomy of retrocaval ureter and associated CVA is troublesome on not only urological surgery but also abdominal lymphadenectomy.

Congenital IVC anomalies are one of the risk factors of in the development of deep venous thrombosis [10, 22]. Thrombophlebitis in deep venous thrombosis might be the cause of stenosis and adhesion between retrocaval ureter and IVC. Asymptomatic retrocaval ureter without hydronephrosis is incidentally detected in CT scans. Even though this is a rare condition, high quality CT images with thin slice data reveal the anomaly in detail: CT urography is especially effective (Figs. 1, 4). Understanding of the retrocaval ureter and associated CVA is important, and radiologists should be pointed out the exact anatomy.

Limitations of the study
Our study has some limitations. First, the sample size was too small to evaluate associated diseases. Second, all cases could not be evaluated using CT urography. High loop retrocaval ureter might not be detected at the delayed phase on contrast-enhanced CT. Furthermore, a large study including long-term follow up is necessary to evaluate congenital and acquired disease.

CONCLUSIONS
We identified the prevalence of associated CVA and CRA in patients with retrocaval ureter using CT
data was about 20%. Various CVA with retrocaval ureter were found resulting from abnormal development of not only the right posterior cardinal vein but also other cardinal veins.

Acknowledgements
Authors thank Dr. Shuichi Kawada and Dr. Hiroshi Yamamuro for their technical support.

Conflict of interest: None declared

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