CASE REPORT

Retrocaval Ureter Detected on CT Urography: A Case Report

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Abstract

Retrocaval ureter is a rare congenital anomaly resulting from abnormal embryological development of the inferior vena cava rather than a primary abnormality of the ureter. Although congenital, patients often present in adulthood with symptoms related to upper urinary tract obstruction. A 47-year-old man presented with recurrent intermittent right flank pain, typically precipitated by consuming a large volume of alcohol. Physical examination and laboratory investigations were unremarkable. Ultrasonography demonstrated mild right hydronephrosis and proximal hydroureter without an identifiable cause. Contrast-enhanced CT urography with three-dimensional reconstruction revealed a Type I retrocaval ureter, demonstrating the characteristic fish-hook deformity with the proximal ureter coursing posterior to the inferior vena cava. The patient underwent open ureteroureterostomy with double-J stenting and had complete resolution of symptoms and hydronephrosis on follow-up.

This case highlights the importance of considering retrocaval ureter in adults presenting with recurrent unilateral flank pain and unexplained hydronephrosis. CT urography with multiplanar and three-dimensional reconstructions is the imaging modality of choice for accurate diagnosis, anatomical assessment, and preoperative planning. The case also illustrates the close relationship between embryological development, anatomical variation, and radiological anatomy.

Keywords: retrocaval ureter; circumcaval ureter; hydronephrosis; computed tomography urography; inferior vena cava.

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Introduction

A retrocaval ureter, more accurately referred to as a circumcaval ureter, is a rare congenital anomaly where the proximal ureter deviates medially and travels posterior to the Inferior Vena Cava (IVC). This condition, which occurs more frequently on the right, is caused by the abnormal persistence of the right posterior subcardinal vein and failure of the right supracardinal vein to develop or regress (1, 2). It has an occurrence rate of 1 in 1000 individuals and is predominantly found in males (3). Despite the congenital nature of the anomaly, patients become symptomatic in the 3rd or 4th decade of life. Common symptoms such as flank pain and hematuria arise from hydronephrosis caused by obstruction of the pelvi-ureteric junction, secondary to an adynamic retrocaval segment, compression of the retrocaval segment, or a ureteral kink (4).

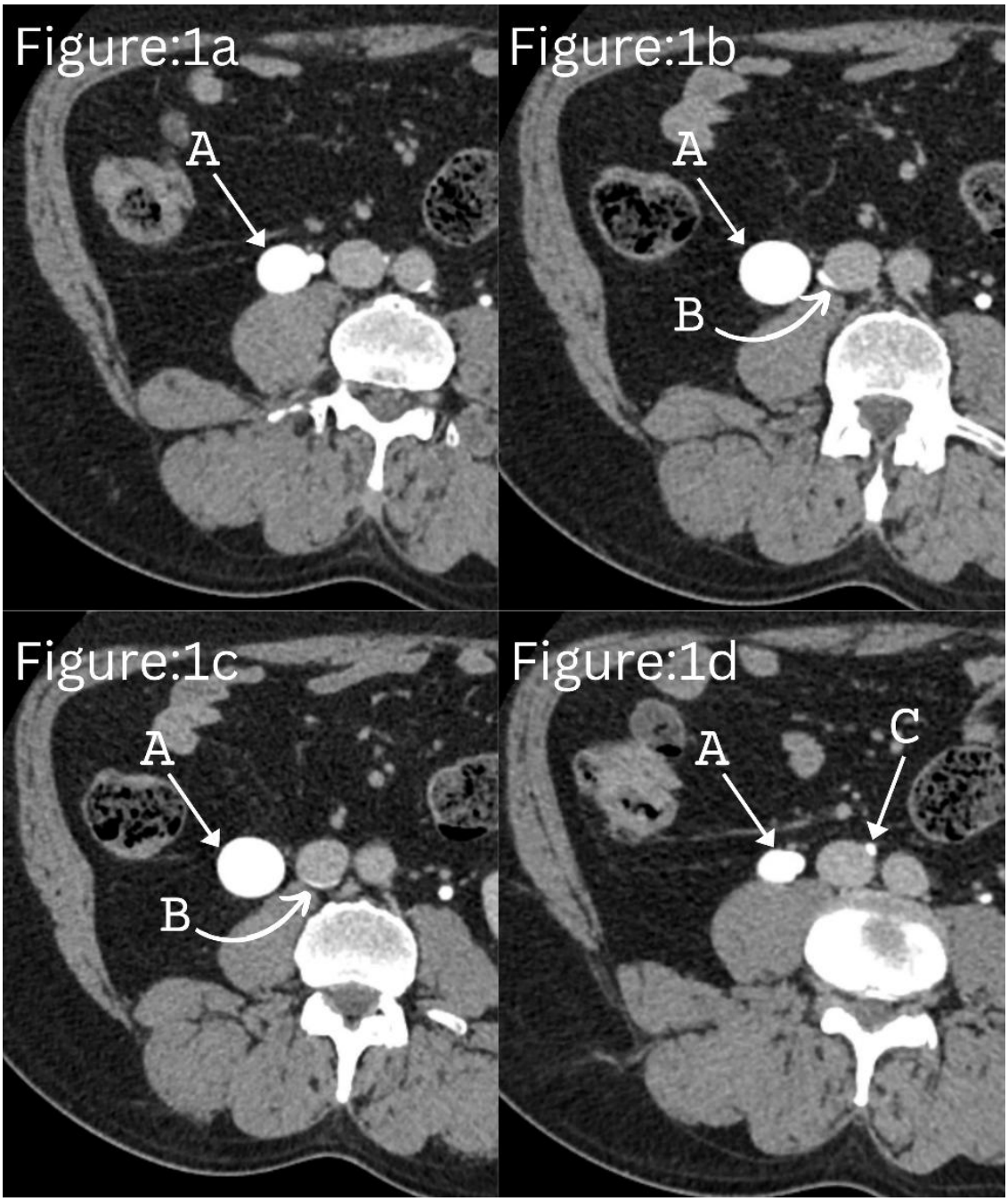
Case Presentation

We report a case of a 47-year-old male who presented to a general practitioner with dull right flank pain after attending a social gathering where he consumed alcohol. He experienced similar flank pain for the last five years, mainly after consuming alcohol. However, he did not seek medical advice until the current episode. The patient denied a history of vomiting, fever, dysuria, hematuria or weight loss. Physical examination was unremarkable except for mild tenderness at the right renal angle. Kidneys were not ballotable. Laboratory investigations, including a full urine report, blood urea, creatinine, electrolytes, complete blood count and C-reactive protein, were within normal limits. Because of unremarkable physical signs, lab investigations, and the history of several similar episodes, he was referred for an ultrasound investigation.

Abdomino-pelvic ultrasound with a full bladder revealed a prominent right renal pelvis with dilated proximal ureter and a dilatation of calyces. Renal cortical thickness and echogenicity were normal. The rest of the ureter was not detected due to overlying bowel gas. No calyceal, renal pelvic, vesicoureteric junction or bladder calculi were detected. An abdominal ultrasound did not reveal a cause for the dilated proximal ureter. A mid-ureteric stricture or calculus was suspected, and the patient was referred for a CT urogram.

The nephrogenic and delayed phases of the CT urogram (CTU) demonstrated mild right-sided hydronephrosis and moderate proximal hydroureter. Three-dimensional (3-D) reconstruction of the CTU showed that the right mid ureter passed behind the IVC and exited medially between the IVC and the aorta, returning to its normal position at the level of the upper border of the L-4 vertebra. At the level of the upper border of the L-4 vertebra, the right proximal ureter passes posterior to the IVC. It exists medially between the cava and the aorta to return to its normal position. At this point, abrupt narrowing of the proximal ureter gives rise to the classic “fish-hook” or “S-shaped appearance”. The distal third of the ureter was of a normal diameter. (Figures 1, Figure 2 and Figure 3) The right ureter joins the normal position of the bladder. No high-density opacities were identified in non-contract CT within the ureteric pathways to suggest ureteric calculi. Based on the CTU with 3-D reconstruction, the final diagnosis was made as a right-sided retrocaval ureter.

The patient underwent open ureteroureterostomy to correct the anatomical variation after obtaining informed consent, followed by insertion of a double-J stent. The double J stent was removed after 6 weeks. He had resolution of the right flank pain and hydronephrosis during follow-up ultrasonography.



Figures 1: Axial CT urography images demonstrate a right retrocaval ureter.
Consecutive axial contrast-enhanced CT urography images demonstrate the abnormal course of the right ureter. (A) Dilated proximal right ureter. (B) The mid ureter passes posterior to the inferior vena cava (IVC). (C) Ureter emerges medially between the inferior vena cava and the abdominal aorta before returning to its normal anatomical course. Moderate proximal hydroureter is evident proximal to the retrocaval segment.

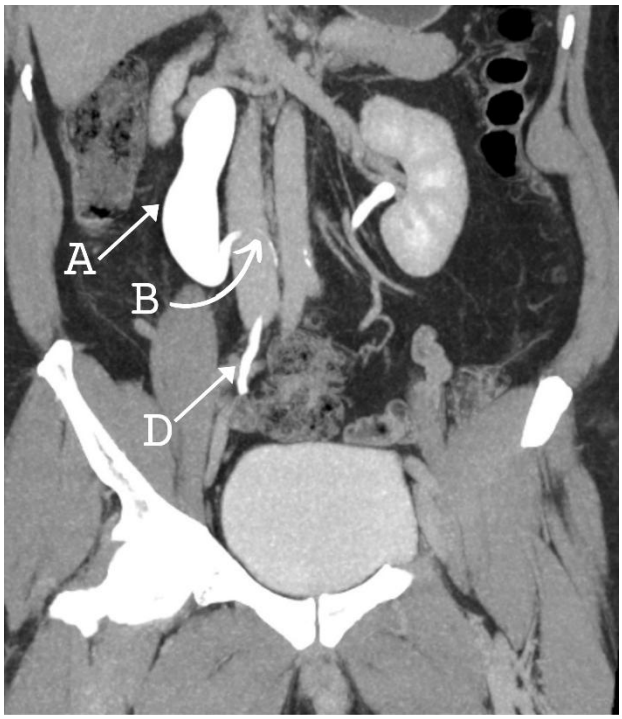


Figure 2: Coronal CT urography images demonstrate the anatomical course of the right retrocaval ureter.
Coronal contrast-enhanced CT urography images demonstrate (A) the dilated proximal right ureter, (B) the retrocaval segment of the mid ureter passing posterior to the inferior vena cava, and (D) the distal ureter returning to its normal calibre and anatomical course inferior to the retrocaval segment.

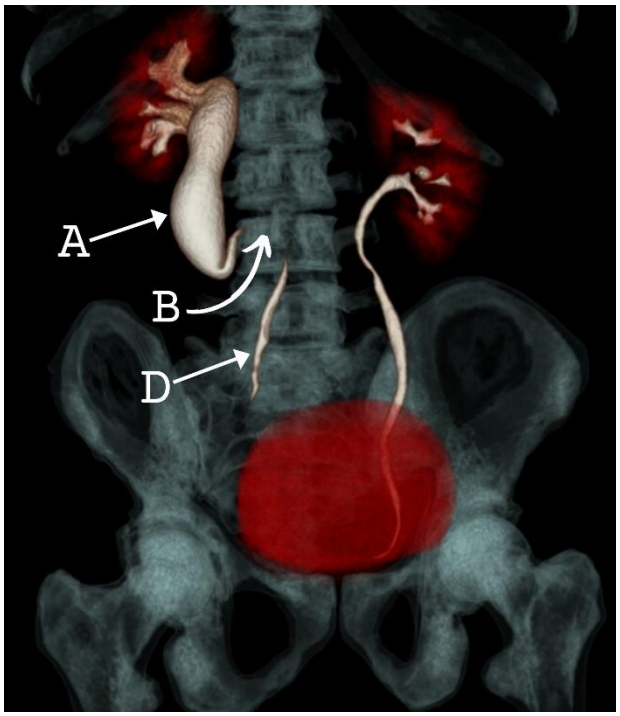


Figure 3: CT urography with three-dimensional reconstruction of the right retrocaval ureter.
Three-dimensional volume-rendered CT urography clearly demonstrates the abnormal anatomical relationship between the ureter and the inferior vena cava. (A) Dilated proximal right ureter. (B) The retrocaval segment of the ureter courses posterior to the inferior vena cava. (D) Distal ureter returns to its normal calibre and anatomical position after emerging anterior to the inferior vena cava.

Discussion

This rarely diagnosed condition, characterised by male predominance, is an embryological venous system anomaly. It predominantly involves the right ureter, with left ureter involvement being associated with situs inversus (3).

The ureter develops from the ureteric bud, an outgrowth of the caudal mesonephric (Wolffian) duct. The stalk of the ureteric bud forms the definitive ureter. Its distal end forms the renal pelvis, calyces, and the collecting ducts of the kidney (5).

In normal development, the right ureter descends vertically from the renal pelvis and anterior to
the psoas major muscle. It remains lateral to the inferior vena cava throughout its abdominal course (6). The ureter crosses the pelvic brim anterior to the bifurcation of the common iliac vessels before entering the true pelvis and finally opening into the urinary bladder (6).

The embryological development of the inferior vena cava (IVC) is a complex process. The adult IVC consists of four embryologic

segments: hepatic, suprarenal, renal, and infrarenal segments (5). Retrocaval ureter is therefore not a primary anomaly of the ureter but is due to abnormal development of the IVC. Persistence of the right posterior cardinal vein as the infrarenal segment of the IVC results in the ureter being trapped posterior to the IVC and not lateral to it, giving rise to the characteristic retrocaval course (5-7). (Figure 4)

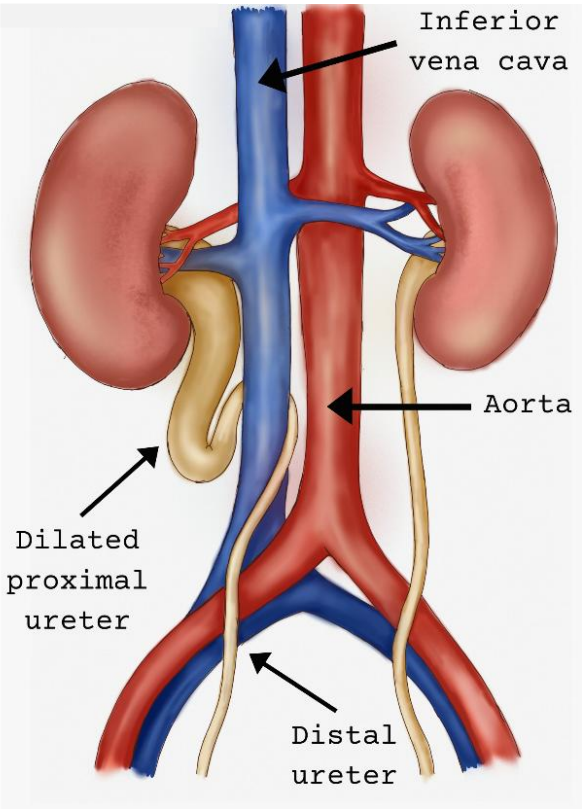


Figure 4: Schematic illustration demonstrating the anatomical relationships of the right retrocaval ureter.
The proximal ureter is dilated above the retrocaval segment and courses posterior to the inferior vena cava before emerging medially between the inferior vena cava and the abdominal aorta, ultimately returning to its normal anatomical position as a distal ureter of normal calibre. (Original schematic illustration by K. P. Imesh Navinda.)

Other differential diagnoses for adult males presenting with unilateral flank pain with imaging features of hydronephrosis and hydroureter include ureteric calculi causing dilatation of the proximal ureter, ureteric strictures, normal variants involving crossing vessels and external ureteric compressions like retroperitoneal fibrosis (8).

Due to the nonspecific symptoms and the rarity of the condition, it may take considerable time to arrive at a proper diagnosis. The condition may progress into complicated right-sided obstructive uropathy and may deteriorate renal function due to progressive back-pressure effects. In addition, this may be a cause for recurrent urinary tract infections and precipitate the formation of stones in the urinary tract (9).

Retrocaval ureter is most often diagnosed as an incidental finding during radiological investigations performed to detect other pathologies. However, most affected individuals are asymptomatic. When symptoms are present, the most common clinical presentation is intermittent right flank pain, usually due to partial upper urinary tract obstruction. A large fluid intake, combined with the mild diuretic effect of alcohol, may transiently increase renal pelvic pressure in patients with partial urinary outflow obstruction, thus precipitating episodic flank pain due to increased urine production. Similar intermittent symptoms have also been described in patients with ureteropelvic junction obstruction.

In the present case, physical examination and laboratory investigations were largely unremarkable. At the same time, the patient's symptoms were intermittent and consistently precipitated by consuming a large volume of beer. CTU with 3-D reconstruction subsequently demonstrated a retrocaval ureter with mild hydronephrosis and proximal hydroureter. This relatively subtle and episodic

clinical presentation points out the importance of considering retrocaval ureter as the differential diagnosis of adults presenting with recurrent unilateral flank pain and unexplained hydronephrosis, especially when no ureteric calculus or other obvious cause of obstruction is identified on imaging.

Retrocaval ureters are categorised as Type I and Type II, with Type I being the most common, accounting for more than 90% of reported cases (10). Type I is usually symptomatic, and it is associated with moderate to severe hydronephrosis and, in about half of cases, with extreme medial deviation of the middle ureteric segment, giving a classic “S-shaped appearance” or “fishhook” deformity. Type II patients show less medial deviation of the ureter with mild or absent hydronephrosis. It is less common and shows less obstruction, with the proximal ureter crossing the IVC at a higher level horizontally, at the level of the renal pelvis or the uteropelvic junction. In Type II, intravenous urogram (IVU) or CTU will show this as a ‘sickle-shaped’ deformity. A type I retrocaval ureter was observed in our patient, with the 3-D reconstruction of the CTU demonstrating the classic “fishhook” appearance.

Ultrasound of the abdomen is useful as an initial investigation, as with our patient, demonstrating the dilated pelvicalyceal system and proximal ureter without evidence of ureteric calculi. However, ultrasound may not adequately visualise the ureter passing posterior to the IVC. CTU with 3-D reconstruction is considered the imaging modality of choice, as it clearly demonstrates the abnormal retrocaval course of the ureter. The course of the right ureter is also clearly visible, with the mid ureter passing posterior to the IVC and exiting medially between the inferior vena cava and aorta to return to its normal course. It will also demonstrate the "fishhook" appearance seen in Type I and the sickle shape seen in Type II. MRI urography (MRU) provides a useful radiation-free alternative, specifically in younger patients and in those with contraindications to iodinated contrast. In the case reported by Igbokwe *et al.,* CTU successfully documented the characteristic "fish-hook" deformity. However, the distal ureter was not opacified on delayed images, illustrating that complete delineation of the ureter may occasionally be limited with CTU alone (11).

Knowledge of the characteristic radiological signs of a retrocaval ureter is important because the diagnosis depends almost entirely on radiological imaging. Recognition of the classic "fish-hook" deformity in Type I or the "sickle-shaped" appearance in Type II is essential to exclude other causes of upper urinary tract obstruction. Accurate radiological assessment is also fundamental for preoperative planning, as it clearly demonstrates the abnormal relationship between the ureter and the inferior vena cava. Multiplanar CT reconstruction with 3-D imaging provides excellent visualisation of the entire ureteric course and its characteristic anatomical features, facilitating both diagnosis and surgical planning.

Conservative management with regular clinical and radiological follow-up is adequate for asymptomatic patients without significant obstruction or deterioration of renal function. Surgical intervention is suggested for symptomatic patients with progressive hydronephrosis, impaired renal function or recurrent urinary tract infections. The usual surgical management is ureteroureterostomy, which aims to reposition the ureter anterior to the IVC and relieve the ureteric obstruction. This is achieved by dividing the ureter, transposing it anterior to the IVC, and

performing a tension-free ureteroureterostomy or pyeloureterostomy over a double-J stent. The retrocaval segment, which may be stenotic or aperistaltic, may be excised or preserved depending on the intraoperative findings and surgeon preference (12).

Traditionally, this procedure was performed through an open approach. However, laparoscopic and robot-assisted techniques have gained significant popularity because they provide the same surgical outcomes while offering fewer post-surgical complications (3, 12, 13). Accurate preoperative radiological imaging is essential to identify the abnormal anatomical relationship between the ureter and the IVC. This is important for surgical planning and to reduce the risk of accidental intraoperative complications.

This case is an excellent example of how embryological development influences adult anatomy and demonstrates the close relationship between vascular development and congenital urological anomalies. This emphasizes how abnormal development of the IVC can alter the normal course of the ureter, leading to upper urinary tract obstruction. Diagnosis of these complex anatomical variations depends on cross-sectional imaging with multiplanar and 3-D reconstructions, which accurately illustrate the relationships between the ureter and surrounding vascular structures. Incorporating such cases into anatomy teaching enhances the understanding of applied embryology and radiological anatomy while increasing awareness of this rare condition among medical students, radiologists, and urologists. This knowledge is invaluable in formulating an accurate differential diagnosis, planning appropriate surgical intervention, and minimising the risk of inadvertent intraoperative injury.

Conclusion

Retrocaval ureter is a rare congenital vascular anomaly, which should be considered in the differential diagnosis of adults presenting with recurrent unilateral flank pain and unexplained hydronephrosis, particularly when a ureteric calculus or other obvious cause of obstruction is absent. Contrast-enhanced CT urography with multiplanar and three-dimensional reconstruction is the modality of choice as it accurately illustrates the characteristic anatomical relationship between the ureter and the inferior vena cava. This allows prompt diagnosis and effective surgical planning. This case presents a strong example of instances of embryological abnormalities and anatomical variations together.

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