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Appearances of the circumcaval ureter on excretory urography and MR urography: A single-center case series

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Abstract

Objectives: To describe Magnetic Resonance Urography (MRU) appearances of the circumcaval ureter, a rare congenital cause of hydronephrosis. **Materials and Methods:** Seven cases of circumcaval ureter, suspected on intravenous urography (IVU), underwent subsequent static MRU using heavily T2-weighted sequences. **Results:** The various appearances of circumcaval ureter on IVU and MRU were studied and compared. The circumcaval portion of the ureter was especially well seen on axial MRU sections, though this portion was routinely not visualized on IVU. In one case with a ureteric calculus, MRU also depicted a circumcaval course of the ureter, thus providing a complete diagnosis. In yet another case, where a circumcaval ureter was suspected on IVU, MRU proved the actual cause of ureteric obstruction to be a crossing vessel. **Conclusion:** Static MRU using heavily T2-weighted coronal and axial sequences can make or exclude the diagnosis of circumcaval ureter unequivocally.

Key words: Circumcaval ureter; hydronephrosis; intravenous urography; magnetic resonance urography

Introduction

Circumcaval ureter is an uncommon congenital cause of obstructive hydronephrosis, caused by the ureter coursing around the inferior vena cava (IVC). The radiological diagnosis of circumcaval ureter has evolved from intravenous urography (IVU), venacavography, and ureteral catherization[1,2] to less-invasive and more informative modalities like Computed Tomographic Urography (CTU) and Magnetic Resonance Urography (MRU).[3] While CTU is admittedly superior in ruling in or out possible common causes of a non-specific urological presentation, MRU is finding its niche in the diagnosis of causes of obstructive hydronephrosis.

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The MRU findings of circumcaval ureter were first described in a case in 2002[3] and more recently in a dog.[4] While the diagnosis of circumcaval ureter in an excreting kidney can be made by CTU or in a non-excreting kidney might be suggested by a non-contrast CT, this series of seven cases illustrates the ability of MRU to clearly depict relevant anatomic relationships and provide confirmatory diagnosis without radiation exposure or the use of contrast medium. To the authors' knowledge, this is the largest series describing MRU findings of circumcaval ureter.

Cases and Methods

Over a 2-year period from 2005 to 2007, all patients suspected to have circumcaval ureter on IVU subsequently underwent MRU. The study was approved by the Institute Ethics Committee and informed consent was taken from the patients prior to the procedures. Static MRU was performed in a 1.5 T Magnetom scanner (Siemens, Erlangen, Germany) using a phased-array body coil. Heavily T2-weighted sequences like Half-fourier Acquisition Single-shot Turbo spin Echo (HASTE) and Fast Imaging with Steady-state Precession (TruFISP) in axial and coronal planes, with thin and thick coronal

maximum intensity projection (MIP) reconstructions were employed. Contrast was not administered, and as is our routine in assessing hydronephrotic systems, patients were not required to undergo oral or intravenous hydration prior to the scan.

Case 1

A 34-year-old man presented with intermittent right flank pain for 6 months. Preliminary ultrasound for suspected calculus disease showed right hydroureteronephrosis with a dilated proximal ureter but no evidence of calculi. Excretory urography showed right-sided grade 4 hydroureteronephrosis, the ureter being dilated up to the L4 vertebral level, with a tapered segment seen coursing superiomedially. A diagnosis of circumcaval ureter was suggested, and MRU was performed. MRU showed grade 4 hydronephrosis with the medialized segment of the ureter coursing around the IVC, and axial sections clearly depicted the anatomy [Figure 1].

Case 2

A 20-year-old man presented with intermittent right flank pain for 1 year and dysuria for 1 week. Sonography revealed right-sided hydronephrosis, and excretory urography showed a mid-ureteric medialization with grade 4 hydronephrosis and the rest of the ureter was not visualized. MRU was subsequently performed, which showed the right upper ureter conically dilated and tapering into a normal caliber ureter that made an S-shaped curve around the IVC.

Case 3

A 12-year-old boy was incidentally found to have right hydronephrosis on routine sonography, and the left kidney was not visualized. An excretory urogram showed grade 3 hydronephrosis on the right, with a dilated proximal ureter and a short distal medialized segment. There was no excretion on the left side. MRU confirmed a solitary right kidney with grade 3 hydronephrosis, and the acute medialization was seen to be due to the ureter winding around the IVC at L3 vertebral level. The patient, being asymptomatic, was not operated, and is on regular sonographic follow-up.

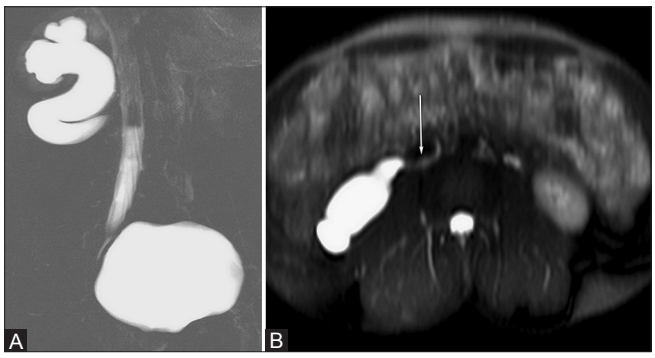

Figure 1 (A, B): MRU appearance of circumcaval ureter. (A) The "sea horse" appearance of a type 1 circumcaval ureter seen on thick slab T2 turbo spin echo coronal MRU image. (B) axial T2-weighted HASTE MRU depicting the ureter coursing around the IVC

Case 4

A 40-year-old woman presented with right flank pain and fever for 2 weeks. Ultrasound showed right hydronephrosis with moving internal echoes, suggesting pyonephrosis. Urine culture was positive for gram-negative bacilli. Excretory urography done after treatment of the infection showed right-sided grade 3 hydronephrosis with a "reverse J" configuration of the dilated system. MRU images and MIP reconstructions showed a similar appearance, but axial sections clearly depicted a circumcaval course of the ureter.

Case 5

A 25-year-old man presented to the emergency department with acute severe right flank pain for a day and history of similar milder pain for the last year. Ultrasound revealed right hydroureteronephrosis with proximal ureteric calculus. IVU showed right grade 4 hydronephrosis with a calculus overlying the pedicle of L3 vertebra. CT showed right hydronephrosis with renal and proximal ureteric calculi, but the intimate relation of ureter and IVC raised the possibility of a circumcaval ureter. MRU unequivocally depicted the circumcaval course of the ureter and two calculi proximal to the obstruction [Figure 2].

Case 6

A 41-year-old man presented with dysuria of 6 months duration. Ultrasound showed right-sided hydronephrosis with a prominent pelvis, the ureter not traceable. IVU showed right grade 2 hydronephrosis, and the "reverse J" shape of the collecting system suggested circumcaval course of the ureter. MRU showed grade 2 obstruction due to hooking of the proximal ureter around the IVC [Figure 3]. This was well made out on both coronal MIP images and axial source images.

Case 7

A 43-year-old man presented with flank pain for 2 months. Sonographically, there was right-sided hydronephrosis. IVU showed a dilated pelvis and proximal ureter, with grade 3 hydronephrosis. Retrograde Pyelogram (RGP) was performed, which showed only the distal normal ureter, up to L4 vertebral level, the tip of the catheter being more medial than expected, causing circumcaval ureter to enter the differential. CT was performed to exclude radiolucent calculus, and the proximity of the ureter and IVC was noted, though there was no retrocaval segment. MRU was performed, and though the procedure had to be terminated prematurely owing to the patient's claustrophobia, the obtained sections excluded a circumcaval course of the ureter and were suggestive instead of a crossing vessel [Figure 4]. The latter was proven intraoperatively a week later.

Discussion

Circumcaval ureter, which has also been called "retrocaval" ureter, is a rare congenital anomaly, which is found in

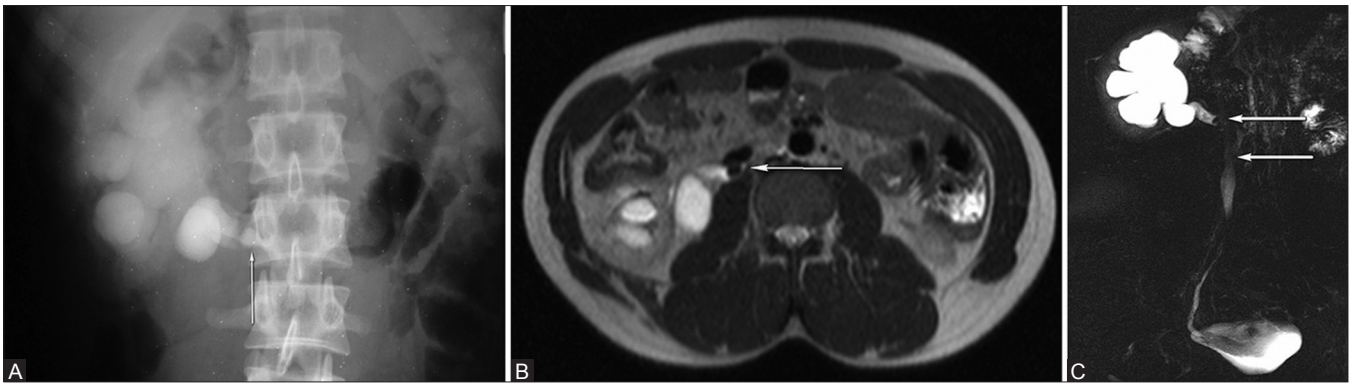

Figure 2 (A-C): Circumcaval ureter with calculus in the proximal obstructed ureter. (A) IVU showing grade-4 hydronephrosis and an opacity adjacent to L3 vertebral pedicle (arrow). (B) Axial MRU section clearly showing the ureteric segment posterior to the IVC with a signal void of a calculus in that segment (arrow). (C) T2-turbo spin echo coronal thick slab MRU showing the calculus as a signal void (upper arrow) and the circumcaval course of the ureter (lower arrow)

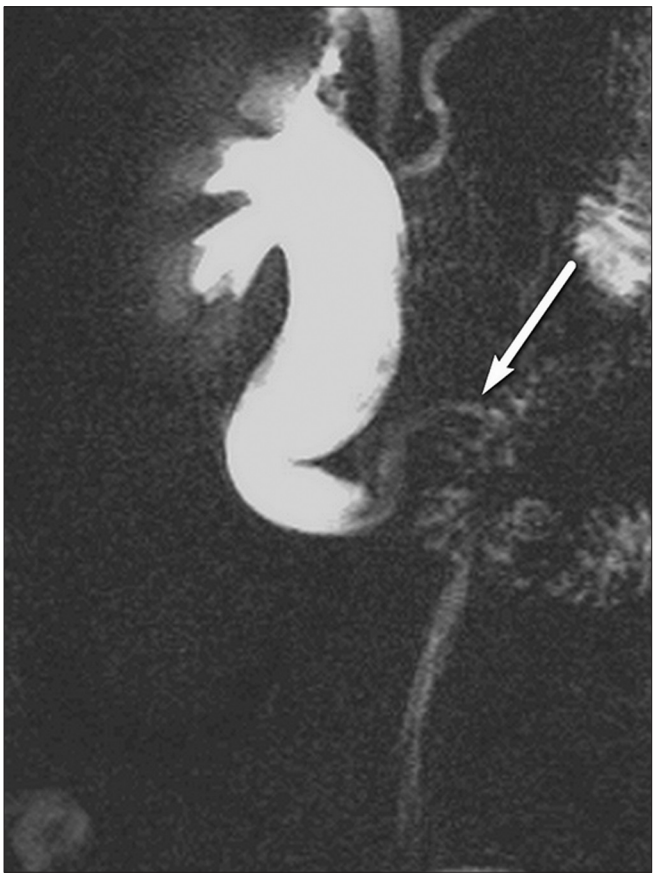

Figure 3: Grade-2 hydronephrosis with circumcaval ureter seen on thick slab T2-turbo spin echo coronal MRU image. The hooking of the proximal ureteric segment can be appreciated (arrow)

about 1 in 1100 cadavers.[5] Males are more often involved than females, with a 2.8:1 ratio. This abnormality is traced embryologically to the anomalous development of the infrarenal IVC from the ventrally located right posterior cardinal vein rather than from the supracardinal vein which is dorsal to the ureter.[6] Expectedly, circumcaval ureters are predominantly right sided. Left-sided circumcaval ureter has been reported in association with a left-sided IVC,[7] as well as in situs inversus,[8] and there is also a report of bilateral circumcaval ureters in association with IVC duplication.[9]

Many anatomic variants of the circumcaval ureter have been described, but radiologically there are two types.[10] Type 1, the common type, shows moderate to severe hydronephrosis with extreme ureteral medialization, usually beyond the pedicle at L3 vertebral level. The ureter shows the classically described "reverse J" or "fish hook" or "sea horse" appearance. In type 2, there is milder obstruction, with less medialization of the ureter, and a "sickle-shaped" configuration is characteristic.

The diagnosis of circumcaval ureter can be suggested on IVU, which demonstrates the medialization of the ureter at L3 or L4 vertebral level with sharp hooking toward the pedicle. Rarely, there may not be obstruction, or the obstruction may be of a lower grade, when the diagnosis might be overlooked. The distal ureter is usually not visualized on IVU, and in some cases, other causes of medialization of the ureter cannot be adequately excluded.[2]

An RGP is usually the next step. Inferior venography, though rarely performed today, has also been used in the past to strengthen the diagnosis.[6] Contrast-enhanced CT with delayed images has been found to be able to depict the ureteral relations to the IVC, and unlike earlier methods, can also adequately exclude extrinsic compression or displacement of the ureter.[2,3] Like IVU, this also requires adequate excretion by the kidney, which might not be present in a chronically obstructed system.

The MRU appearance of circumcaval ureter was first described in 2002,[1] and several studies have shown the diagnostic reliability of heavily T2-weighted sequences in depiction of an obstructed system.[9,10] While coronal MIP images provide the closest similarity to an IVU film,

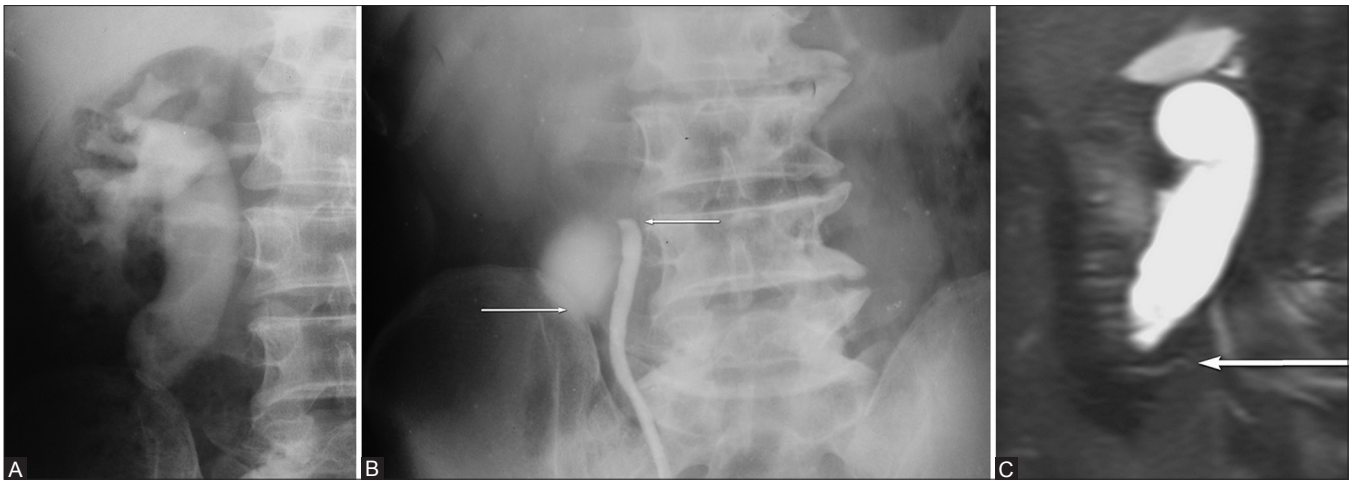

Figure 4 (A-C): Crossing vessel causing ureteric compression. (A) IVU showing grade-2 to -3 hydronephrosis with a grossly dilated pelvi-urteric segment and no cause for obstruction. (B) retrograde pyelography showing the catheter tip (upper arrow) to be superior and medial to the obstructed ureter (lower arrow) suggesting that this segment might be hooked around the IVC. (C) coronal MRU section showing a vessel (arrow) crossing at the site of ureteric narrowing, yielding the diagnosis

it is the axial sections that we found to be most useful in confirming the circumcaval course of the ureter. Indeed, while coronal images often suffered from bowel and other image degrading artifacts, the latter did not hamper tracing the path of the ureter in axial sections. The intrinsic contrast of urine in heavily T2-weighted images enabled easy diagnosis irrespective of excretory ability of the kidney. Static MRU using heavily T2-weighted images is, however, prone to flow artifacts misdiagnosed as filling defects, and the bright signal of fluid is known to obscure small calculi,[11] which require a CT for a definitive diagnosis.

In our series, while cases 1, 2, 4, 5, and 6 showed the "reverse J" appearance described as type 1 circumcaval ureter, case 3 showed a more gradual tapering and less medialization, characteristic of type 2 circumcaval ureter. While the diagnosis was suspected in most of the cases by IVU, case 5 might have been misconstrued as obstruction due to ureteral calculus if not for the medial location of the calculus, overlying the pedicle. On IVU, a medially directed short segment of ureter of normal diameter immediately distal to the dilated segment was a good pointer to the diagnosis. However, this finding is not always present, or may be very subtle, as seen in case 3. Grades of hydronephrosis varied from grade 4 in cases 1, 2, and 5; grade 3 in cases 3, 4, and 7; and grade 2 in case 6, raising the possibility that though a congenital condition, the onset of obstruction might be delayed. One explanation of the fewer reports of this condition in literature than its incidence could thus be the absence of symptomatic obstruction.

Pyonephrosis as seen in case 4 and calculi as in case 5 are two of the potential complications of circumcaval ureter.[7] While the infection in case 4 subsided with antibiotics, the calculi in case 5 required a pyelolithotomy prior to curative surgery.

Conclusions

The diagnosis of circumcaval ureter, so far in the domain of IVU and other invasive methods, can be readily made with static MRU. While circumcaval ureter can have a variety of appearances and grades of hydronephrosis on IVU, the diagnosis can be confidently made or excluded with MRU, as can other causes of obstruction be suggested. The absence of contrast usage, lack of ionizing radiation, and multiplanar imaging capability make MRU an attractive option in the workup of suspected cases where these would want to be avoided, like in the pediatric population, in patients requiring long-term follow-up, in pregnancy, renal failure, non-excreting renal systems, and to corroborate or confirm doubtful CT findings. This series shows the particular usefulness of axial sections in depicting the abnormal ureteric course, even if standard coronal images and reconstructions are equivocal. Valuable anatomical and pathological information can be obtained to guide surgeons and thus affect outcome.

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