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Case Report

Radiology Section

An Intriguing Case of Circumcaval Ureter

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ABSTRACT

Circumcaval ureter is a rare congenital condition resulting from the anomalous development of inferior vena cava due to developmental failure of supracardinal system. This results in posterior and medial looping of the right ureter around the developed IVC.

Keywords: Fish hook deformity, Medial displacement of ureter, Retrocaval ureter, Supracardinal vein, Upper hydroureteronephrosis.

CASE REPORT

A 38-year-old adult male presented with on and off pain in right loin region, since 4-5 years. The pain was vague, not radiating, not related to inspiration or food intake. Patient also gave recurrent episodes of fever. Clinical examination was normal.

Patient was referred for an ultrasound examination to evaluate the cause for his recurring right abdominal pain.

Ultrasound examination showed mild pelvicalyceal dilatation. Upper ureter showed mild dilatation and appeared to pass medial to inferior vena cava. Distal course of ureter could not be traced.

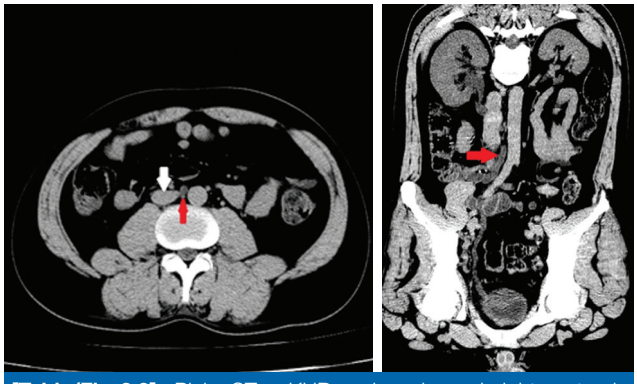
Further evaluation with Intravenous Urography (IVU), Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) was performed.

IVU showed mild hydronephrosis on the right side. Upper ureter was dilated and showed upper ureter taking a medial course up to the level of L3 and then a downward course along the bodies of the vertebral bodies up to L4 and then along the sacral body and then inserting into the bladder [Table/Fig-1].

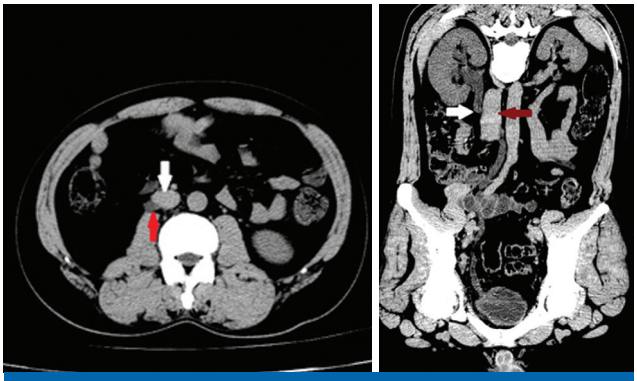
Plain CT of KUB region showed same findings as IVU and further the ureter at the level of L3 is seen posterior to the IVC [Table/Fig-2,3] and then is seen between IVC and aorta upto L4 level [Table/Fig-4] and later is seen anterior to common iliac vein [Table/Fig-5] and then in its normal course upto the bladder.

MRI showed same findings as CT. Course of Ureter crossing posterior to the IVC at level of L2 [Table/Fig-6] and then hooking back to its normal course at level of L4 anterior to common iliac vein on the right side [Table/Fig-7].

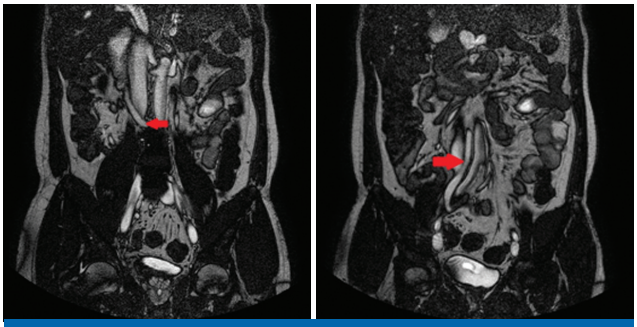
[Table/Fig-1]: IVU shows abnormal medial course of the ureter upto the level of L4 vertebral body with mild proximal hydronephrosis with normal course and insertion of the right ureter into bladder.



[Table/Fig-2,3]: Plain CT - KUB region showed right ureter is in midline between IVC and Aorta in axial section. The same corresponding coronal section showed the ureter being posterior and medial to the IVC at the level of L3 vertebrae.



[Table/Fig-4,5]: The same relation between of right ureter between IVC and Aorta is maintained upto L4 level and later is seen anterior to common iliac vein in axial and coronal sections of plain study. It continues its normal course upto the bladder.



[Table/Fig-6]: T2WI MRI coronal section shows the course of right ureter crossing posterior to the IVC at level of L2 vertebrae.
[Table/Fig-7]: T2WI MRI coronal section shows hooking of the ureter back to its normal course at level of L4 vertebrae anterior to common iliac vein on the right side.

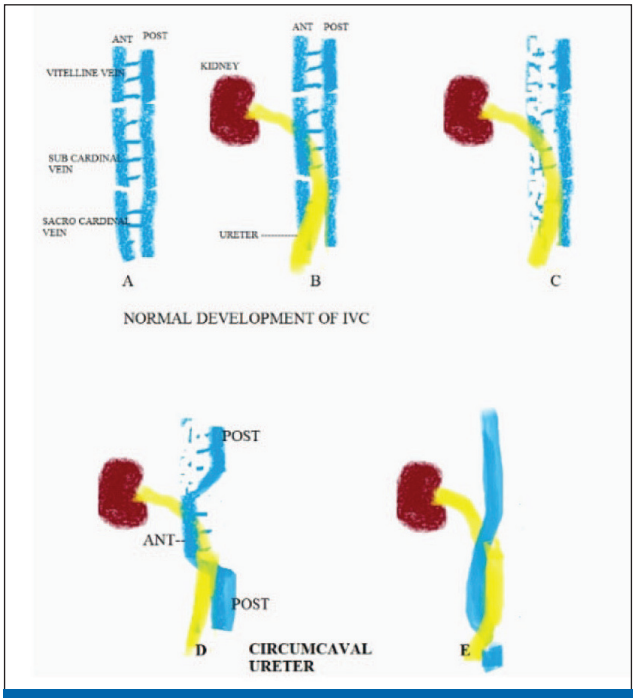
Based on clinical features and radiological findings, we arrived at the diagnosis of Circumcaval ureter. Patient was treated medically with pain killers and was followed up for 1 year. As the patient was asymptomatic, no surgical intervention was carried out.

DISCUSSION

Circumcaval ureter is a rare developmental anomaly of inferior vena cava and not to be mistaken for developmental defect in ureter with incidence of 1-3% with male predominance (2.8 times more commonly in males than females) [1]. Circumcaval ureters have been classified into two types - "Low loop"/ Type 1 is the more common form which is characterized by "typical S" or "fishhook" deformity of the ureter at the level of the obstruction causing moderate to severe hydronephrosis, with obstruction in the lateral margin of the IVC at the level of the third lumbar vertebra (L3). "High loop" / type 2, ureter has a "sickle-shaped" curve, with the point of obstruction at the lateral margin of the IVC with mild or no hydronephrosis. This second variant is rare, and represents around 10% of the known cases.

EMBRYOLOGY

Normally vitelline vein, subcardinal vein and sacrocardinal vein sequentially undergoes development followed by anastomosis and then regress to finally form the definitive inferior vena cava. Embryological right vitelline vein forms the pre-renal segment, right subcardinal vein forms the renal segment while sacrocardinal vein forms the post renal IVC [Table/Fig-8].During development, if the right posterior cardinal vein in the lumbar portion does not atrophy but persists, then it remains to form the renal segment of the inferior vena cava [2,3]. The resultant renal segment of vena cava, therefore, developed anterior to the ureter. The net effect is that the ureter is drawn around the renal segment of the inferior vena cava thus causing a circumcaval position of ureter.



[Table/Fig-8]: Pictorial diagram depicting the normal embryological development (A,B,C) and the developmental anomaly of IVC leading to circumcaval ureter (D,E).

This developmental anomaly causes hooking of the right ureter over the IVC which in turn can cause obstructive symptoms in the right kidney and collecting system. Patient can have variable presentation from being asymptomatic to intermittent abdominal pain or renal colic.

On radiological imaging also, findings can be misleading. USG can show normal findings or obstructive signs upto the upper ureter. IVU examination will only show abnormal course of the upper ureter and later it returns to its normal course. This can happen in variety of other conditions like retroperitoneal fibrosis, mass in retroperitoneum and anomalous course of ureter.

Contrast enhanced CT study will give a more definitive diagnosis of retrocaval ureter. Axial and reconstructed coronal images will show the upper ureter to take a medial course and passing posterior to the IVC and then traversing medial to IVC. Later it hooks around the IVC to come back to its normal course. CT will also demonstrate obstructive signs if any which is usually seen upto the level of hooking of ureter [4]. CECT also will rule out mass lesions and retroperitoneal fibrosis.

MRI will also give the same information as CT. With multiplanar imaging the ureters course in MR imaging will demonstrate the posterior position of ureter in relationship to the IVC in circumcaval ureter. Further, the ureter will be seen hooking back and returning to its normal course [5].

CONCLUSION

When right ureter does not show normal course on USG, CECT and MRI will give more information towards making a diagnosis of circumcaval ureter. MRI is a good alternative to CECT without radiation. Knowing such rarity, helps in clinching the diagnosis with ease among other differentials and alert the clinician accordingly during other urological emergencies which requires further intervention or management.

REFERENCES

[1] Sandercoe GD, Brooke-Cowden GL. Developmental anomaly of the inferior vena cava. *ANZ J Surg*. 2003;73:356–60.
[2] Bass JE, Redwine MD, Kramer LA et al. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics*. 2000; (3):639-52.
[3] Herman T, McAllister W: Radiological clinics of North America. 1991;2(29).
[4] Lautin EM. CT diagnosis of circumcaval ureter. *AJR Am J Roentgenol.* 1988;150(3):591-94.
[5] Uthappa MC, Anthony D, Allen C. Case report: retrocaval ureter: MR appearances. *Br J Radiol*. 2002;75:177-79.

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