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RETROCAVAL/CIRCUMCAVAL URETER: A RARE CONGENITAL ANOMALY

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ABSTRACT
A retrocaval or circumcaval ureter is a rare congenital anomaly which results from the persistence of posterior cardinal vein as the renal segment of the inferior vena cava during embryogenesis. In 1893, Hochstetter first observed the case of retrocaval ureter. Though initially it was thought to be an anomaly in the development of ureter but the studies in embryology have revealed that it is an anomaly related to the development of the inferior vena cava. Review of literature suggest that approximately about 1 in 1000 people have been reported of retrocaval ureter [5] with male to female ratio of 3 : 1. It commonly found on the right side and is usually associated with anomalies of inferior vena cava. In case of situs inversus or in presence of double inferior vena cava retrocaval ureter can be found on the left [6 & 7]. Computed Tomography and Magnetic Resonance studies are the best tools to confirm the diagnosis and to prevent further surgical complications.

CASE REPORT

A 46 year old female patient came in to Emergency Department at Sangre Grande Hospital with complaints of right upper quadrant pain. The pain was dull and intermittent. No abnormality was found on clinical examination, except for minimal tenderness in the right upper quadrant on deep palpation. Complete laboratory evaluation including urinalysis and complete blood picture were within normal limits. Ultrasonography of the abdominal area was performed to rule out for any pathological findings, where she was diagnosed with cholelithiasis. No evidence of cholecystitis. The patient did not have any significant personal or family medical history. The patient was treated and referred to surgical ward. On follow up in the surgical clinic, were an elective cholecystectomy surgery was arranged accordingly. The pre-surgical physical examination and evaluation was unremarkable and the patient underwent a successful laparoscopic cholecystectomy. On post-surgery, patient was experiencing fever and she was referred to the radiology department for a contrast enhanced CT of the abdomen and pelvis to rule out any post-surgical collections.

The CT scan showed small amount of collection in the GB bed in keeping with acute post-surgical findings. However, incidentally there was dilated right renal pelvis and proximal ureter up to the level of L3 where at this point the ureter is narrowed and curves behind the IVC to its medial side and courses down. The findings are in keeping with a right retrocaval ureter [Fig 1 & 2]. On review of the old history and CT done on the same patient the same findings where seen confirming the diagnosis. The rest of the CT scan study was unremarkable.

The patient was managed with uneventful recovery and was discharged from the hospital.

DISCUSSION

The classical course of the ureter begins within the renal sinus as a funnel-shaped dilatation, called the renal pelvis. Pelvis issues from the hilus of the kidney,

descends along its medial margin, or partly behind it. Gradually it narrows till at the lower end of the kidney it becomes the ureter proper. The ureter passes downwards and slightly medially on the psoas major muscle, and enters the pelvis by crossing in front of the terminations of the common iliac artery. In the lesser pelvis the ureter at first runs downwards, and slightly backwards and laterally, following the anterior margin of the greater sciatic notch. Opposite the ischial spine it turns forwards and medially to reach the base of the urinary bladder.

In our present case the ureter rather than passing in its normal course it deviates medially behind the inferior vena cava and winds around the inferior vena cava and crosses in front of it from a medial to a lateral direction, to resume a normal course distally until it reach the bladder. This type of unusal course was first reported by Hochstetter in 1893 as retrocaval ureter. Retrocaval ureter is a rare condition that results from an anomaly in the development of the inferior vena cava [9]. The incidence was reported to be approximately 1 in 1000 people, with male predominance [10]. The anomalous vessel compresses the ureter, causing varying degrees of hydronephrosis. This abnormality occurs as a result of the right supracardinal system failing to develop normally. The right posterior cardinal vein persists and therefore ends up passing infront of the ureter. Normally, the inferior vena cava develops from the vitelline vein, subcardinal and sacrocardinal veins, which has to undergo definite order of anastomosis and regression to become the inferior vena cava. Normally, the right vitelline vein forms the pre-renal or hepatic segment of the inferior vena cava, the right subcardinal vein forms the renal segment and the right sacrocardinal vein forms the post-renal vena cava. The development of retrocaval ureter is due to the abnormal development of the inferior vena cava as a result of failure of degeneration of the right posterior cardinal vein in the lumbar portion [11]. Retrocaval ureter is usual present on the right side and rarely it can be seen on the left side when it is associated with situs inversus or double inferior vena cava [12&13]. The commonest presenting complaint is loin pain caused by hydronephrosis due to compression of the right ureter between the inferior vena cava and psoas muscle as ureter courses posterior to inferior vena cava.

Based on the radiological appearance of retrocaval ureter in relation to inferior vena cava, are divided into two types. In Type 1, the ureter crosses behind the inferior vena cava and has an S-shaped deformity of the ureter. Marked hydronephrosis is present in over 50% of patients in this type. In Type 2, the retrocaval segment is seen at the level of the renal pelvis in a sickle-shape appearance with mild hydronephrosis and is less common compared with Type 1 [14&15].

Figure 1. CT scan demonstrates a more medial retrocaval placement of the right ureter.

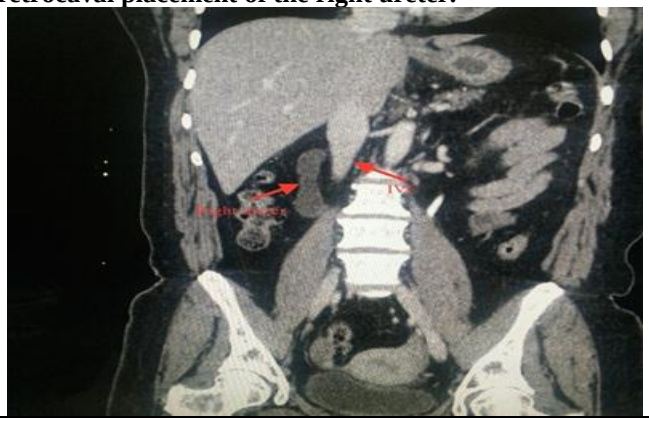
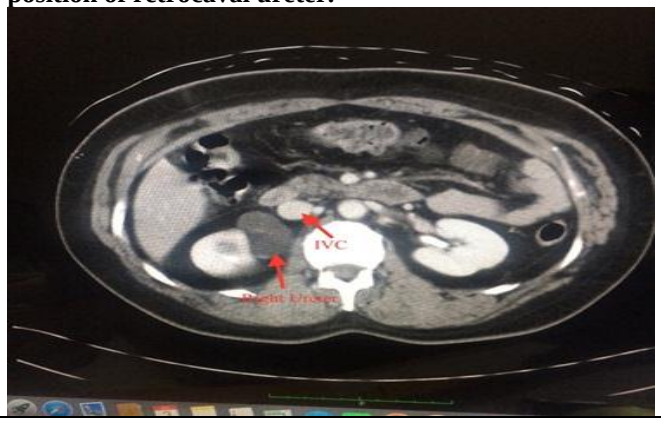


Figure 2. Axial CT scan at the level of L3 showing the position of retrocaval ureter.



CONCLUSION

The objective of this case is to emphasize the importance of diagnosing of retrocaval or circumcaval ureter which might result in misinterpretation of the radiological features. These variations have to be recognized in order to avoid surgical complications when planning for renal transplantation or nephrectomy.

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CONFLICT OF INTEREST: NIL

REFERENCES

1. Hochstetter F. Beitrage zur entwicklungsgeschichte des venen. (1893). Systems der amnioten: III. Sauger Morph Jahrb, 20:542.
2. Dreyfuss W (1959). Anomaly simulating a retrocaval ureter. *J Urol*, 82, 630.
3. Lerman I, Lerman S, Lerman F. Retrocaval ureter (1956). Report of a case. *J Med Soc N J*, 53:74.

4. Peisojovich MR, Lutz SJ. (1969). Retrocaval ureter-a case report and a successful repair with a new surgical technique. Mich Med, 68(21), 1137–1141.
5. S.V.S. Soundappan and A.P. Barker. (2004). Retrocaval ureter in children: a report of two cases. *Pediatric Surgery International*, 20(2), 158–160.
6. Carrion H, J Gatewood, V Politano, G Morillo and C Lynne. (1979). Retrocaval ureter: report of 8 cases and the surgical management. *Journal of Urology*, 121(4), 514–517.
7. Ishitoya S, Y Arai, K Waki, K Okubo and Y Suzuki. (1997). Left retrocaval ureter associated with the Goldenhar syndrome (branchial arch syndrome), The Journal of Urology, 158(2), 572–573.
8. Bass FE, Redwine MD, Kramer LA, Huynh PT, Harris JH. (2000). Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radio Graphics*, 20, 639–652.
9. Lautin EM, Haramati N, Frager D, Friedman AC, Gold K, Kurtz A (1988). CT diagnosis of circumcaval ureter. *AJR Am J Roentgenol*, 150(3), 591–594.
10. Uthappa MC, Anthony D, Allen C (2002). Case report: Retrocaval ureter: MR appearances. *Br J Radio*, 75(890), 177–179.
11. Arianna Lesma, Aldo Bocciardi and Patrizio Rigatti. (2006). Circumcaval ureter: Embryology. *European Urology*, 5(5), 444-448.
12. Watanabe M, Kawamura S, Nakada T. (1991). Left preureteral vena cava (retrocaval or circumcaval ureter) associated with partial situs inversus. *J Urol*, 145, 1047–1048.
13. Rubinstein I, Calvacanti AG, Canalini AF, Freitas MA, Accioly PM. (1999). Left retrocaval ureter associated with inferior vena cava duplication. *J Urol*, 162, 1373–1374.
14. Kenawi MM, Williams DI. Circumcaval ureter. (1976). A report of 4 cases in children with a review of the literature and a new classification. *Br J Urol*, 48, 183–192.
15. Bateson E, Atkinson D. (1969). Circumcaval ureter: a new classification. *Clin Radiol*, 20, 173–177.