Retrocaval ureter - a case report

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Abstract

Retrocaval ureter is a rare congenital anomaly which has an incidence of occurrence of one in 1500 live births. It is known to occur 2.8 times more commonly in males than females. While carrying out a study on congenital anomalies in urinary system in a collection of still born fetuses, a case of retrocaval ureter was noticed in a still born male foetus. In Retrocaval ureter, ureter is S shaped on the right side and a part of inferior vena cava is anterior to the proximal part of the ureter. Here the development of ureter is normal. Whereas the development of inferior vena cava is abnormal.

Key Words: inferior venacava, embryology, congenital anomaly, foetus.

Introduction

A retrocaval ureter is a rare congenital anomaly. It occurs in about one in 1500 live births with two- to threefold male predominance1. Retrocaval ureter also referred to as circumcaval ureter or preureteral vena cava is a rare congenital anomaly with the ureters passing posterior to the inferior vena cava. The ureter classically course medially behind the inferior vena cava winding around it and then passes laterally in front of it to then course distally to the bladder2.

During a study of congenital anomalies in urinary tract in fetuses a case of retrocaval ureter was found in a still born male foetus. The embryological, pathological and clinical significance of retrocaval ureter are discussed in this case report.

Case Report

The retrocaval ureter was discovered in a still born male fetus of 34 weeks. Other wise the head, neck, face, chest and limbs were found to be normal in this fetus. No abnormalities were found during the dissection of the abdomen. Entire gastro intestinal system was found to be normal. Scrotal bags were empty and the testis were found in the inguinal canal (undescended testis).

Following were the findings on examination of urinary system (Fig. 1):

Right kidney was lobulated and measured 3.4 cm in length, 2.4 cm in width and 1.4 cm in thickness. The right hilum was facing medially. The upper pole was at the level of T11 vertebra and lower pole was found at the L2 vertebral level. Length of right ureter was 5 cm which opened into the urinary bladder.

The left kidney was also lobulated and measured 3.3 cm in length, 2.4 cm in width and 1.3 cm in thickness. Here also the hilum was medially placed. Vertebral level of the upper pole corresponded to T11 vertebra and the lower pole to L2 vertebra. Length of the left ureter was 4.9 cm, opening into the urinary bladder.

Right sided ureter was found to be placed posterior to IVC (retro caval). IVC coursed anterior to right renal pelvis and proximal part of the right ureter. (Right testicular vein drained into this segment of vena cava).

Lower down the inferior vena cava was found to lie lateral to the right ureter and then posterior to it, at the level of L3 vertebra. Distal to this, right ureter passed lateral to inferior vena cava and entered the pelvis.

Discussion

A circumcaval ureter is a rare congenital anomaly usually associated with upper urinary tract stasis and an "S" or "fishhook" deformity of the ureter, in which the ureter itself passes behind the inferior vena cava3. Retrocaval ureter is an uncommon embryological anomaly involving the the ureter coursing behind and being obstructed by the inferior vena cava. A review of the literature up to 1961 by De Gironcoli quoted 138 cases, and a further review till 1966 revealed an
additional 74 cases. More than 200 cases have been reported till date. The first observed case of retrocaval ureter was described by Hochstetter in 1893. Though initially thought of as an anomaly of ureteric development studies in embryology has revealed an anomaly related to the development of the inferior vena cava.

**Embryological basis:** A circumcaval ureter results from the posterior cardinal vein persisting as the renal segment of the inferior vena cava during development. Normally, the inferior vena cava develops from the vitelline vein, subcardinal and sacrocardinal veins, which must undergo sequential development, 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. Typically, the circumcaval ureter aetiology is assumed to be abnormal embryologic development of the vena cava as a result of failure of atrophy of the right posterior cardinal vein in the lumbar portion. When the renal segment of inferior vena cava is formed from the right posterior cardinal vein that lies ventral to the ureter, then the ureter will develop in a 'circumcaval' position.

**Presenting symptoms:** Although the abnormality is congenital, patients seldom report symptoms until the third or fourth decades of life. Common presentations include flank pain (right lumbar pain), renal stone and hydronephrosis, recurrent urinary tract infections and microscopic or gross haematuria.

**Diagnosis:** Retrograde ureteropyelogram is considered as not a absolute necessity since Spiral CT scan can detect the ureter and inferior vena cava anomalies and is considered an investigation of choice. MRI can demonstrate the course of a pre-ureteral vena cava which may give more detailed picture and is of advantage in being less invasive imaging modality, without exposure to radiation, when compared with CT and retrograde ureteropyelography.

**Radiological findings:** Ureters normally course about the width of thumb lateral to the lumbar vertebral pedicles and about the width of two fingers medial to pelvic brim in true pelvis. With retrocaval ureter right ureter’s course swings medially over pedicle of L3/4, and passes behind IVC. Then exits anteriorly between IVC and aorta returning to its normal position.

**Clinical types of retrocaval ureter:** Bateson and Atkinson analyzed 92 cases and classified retrocaval ureter in to two types on the basis of radiographic criteria:

- Type I, the more common form, has severe or moderate hydronephrosis with extreme medial deviation of the middle ureteral segment that is usually medial to the pedicle or across the midline at the L3 level. An "S" or "fish-hook" deformity is present at the point of obstruction.

- In type II there is mild hydronephrosis and less medial deviation of the ureter. The ureter is sickle shaped at the level of the obstruction. The other entities that may produce medial deviation of the ureter include retroperitoneal fibrosis and a retroperitoneal mass.

**Retrocaval ureter in children:** Although it is a congenital anomaly it normally presents in the third and fourth decade of life. However cases of retrocaval ureter have been reported in patients of younger age groups and children also. In a literature survey we could not come across any reports of retrocaval ureter in fetuses. Hence the present case is reported as a rarity.

**Unusual presentations of retrocaval ureter:** Retrocaval ureter can be associated with Turner’s syndrome. If it involves the left ureter then it is usually associated with either partial or complete situs inversus or duplication of the inferior vena cava. It has also been reported in association with horseshoe kidney and Goldenhar syndrome.

**Surgical correction:** Surgical intervention is indicated in symptomatic cases. Surgery involves the division of the ureter and repositioning it anterior to the inferior vena cava. This may be achieved through an
Fig 1. Shows the abdominal cavity of the fetus with retrocaval ureter on the right side.

**Abbreviations:**

AO- Aorta; IVC- inferior venacava; Lt K-Left kidney; Rt K -Right kidney; Rt Ur- Right ureter.

Retrocaval ureter: An anastomosis between the renal pelvis and the ureter or a uretero-ureteric anastomosis over a double-J stent. The segment behind the inferior vena cava which may have become aperistaltic, it can either be excised or left in situ. Patients with minimal caliceal dilatation and no significant symptoms do not need surgery but need to be followed up. Transperitoneal and retroperitoneal laparoscopic repair of the retrocaval ureter has become common of late, having advantages like shorter hospital stay and early recovery 1,2,6,9.

**Differential diagnosis:** Important differential diagnosis includes retroperitoneal fibrosis and retroperitoneal masses displacing the ureters from its normal course. Abdomino pelvic CT Scan is helpful in excluding these conditions1,2.

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**References**


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