

CASE REPORT

Retrocaval ureter with ureteral carcinoma

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The authors report a rare congenital anomaly, retrocaval ureter (RCU) with carcinoma. The patient had recurrent pyelonephritis, hematuria and renal colic. Urography shows no function on right side. Abdominal ultrasound revealed marked right hydronephrosis. Antegrade ureterography through nephrostomy revealed a RCU. Helical C.T. confirmed the RCU and also revealed a defect in contrast medium in the upper ureter. We performed percutaneous antegrade nephro-ureteroscopy and found carcinoma of the retrocaval ureter. This may be the first case of tumor in the RCU. We did not find any other in the literature. (Fig. 3, Ref. 14.)

Key words: retrocaval ureter, congenital anomalies, carcinoma, obstruction, spiral C.T.

Retrocaval ureter is a rare congenital anomaly. The first case reported was in 1893 by Hochstetter. The retrocaval position of the ureter is the result of an abnormal development of the fetal abdominal venous channels. The incidence in population is approximately 1 to 5 per 5000 cases (Nielsen, 1959; Primus and Pumer, 1991; Chuang et al, 1974). Postmortem series, however, have shown a higher incidence of approximately 9 per 1000 (Fukuoka et al, 1992; Ekstroem and Nilsson, 1959). It is a rare congenital venous system anomaly rather than a urinary system anomaly and usually requires ureteral surgical correction in symptomatic patients (Grosse et al, 1975). The incidence rate is three to four times higher in males than the females (Kenawi and Williams, 1976; Nielsen, 1959). The occurrence frequency of symptomatic obstruction of retrocaval ureter with ureter carcinoma is not known.

Patients with retrocaval ureter commonly suffer from flank pain, macrohematuria (Primus and Pumer, 1991) or recurrent pyelonephritis (Fukuoka et al, 1992). The formation of renal stones may also occur (Ekstroem and Nilsson, 1959). Due to the gradual development of associated hydronephrosis, patients most often develop complications during their forties and fifties (Polascik and Chen, 1998).

The standard treatment of patients with symptomatic RCU is resection of the RCU and end-to-end anastomosis (Brito et al, 1973) or uretero-pyeloanastomosis by Anderson–Hynes (Primus and Pumer, 1991) anterior to the vena cava. In the past 6–8 years many surgeons have used laparoscopic intervention.

Case report

A 52-year-old male was initially evaluated elsewhere due to two months of right epigastric pain. There was a history of urinary tract infection, right flank pain and elevated temperature. Initial evaluation with abdominal ultrasound revealed marked right hydronephrosis. An urography showed no function of the right kidney. From the patient's history: spontaneous passage of small stones after renal colic, pains as well as hematuria suggested ureterolithiasis. Cystoscopy was normal and right ureteroscopy with 8.5 F semi-rigid ureteroscope was unsuccessful and with 7.5 F flexible ureteroscope was also not successful for bleeding. We performed a percutaneous nephrostomy and antegrade pyelography and ureterography. This revealed a RCU (Fig. 1). Helical C.T. confirmed the RCU (Fig. 2) and also revealed defect in contrast medium in the upper ureter. Next step was nephro-ureteroscopy through nephrostomy and biopsy of suspicious tissue for histopathology examination. The histopathology examination showed ureteral carcinoma (Fig. 3) The left kidney was of

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Fig. 1. Pyeloureterography through a nephrostomy.

normal size and showed good excretion of contrast medium. We performed ureteronephrectomy with a bladder cuff. The pathologist did not find any other tumor in ureter. The patient is now 18 months after operation in good condition, without evidence of TU.

Discussion

The retrocaval position of the ureter is the result of an abnormal development of the fetal abdominal venous channels. The infrarenal part of the inferior vena cava originates from the right supracardinal vein, which runs dorsally to the ureter, thus allowing the ureter to lie lateral to the inferior vena cava. In this particular anomaly, the more ventral infrarenal subcardinal vein persists, thereby obliging the more dorsal placed ureter to pass behind and around it as the developing metanephros ascends from the pelvis to its final lumbar position (Chuang et al, 1974).

There are 2 anatomic types of RCU (Primus and Pumer, 1991). In one, the upper ureter and renal pelvis are almost horizontal as they pass behind the vena cava, there is generally no obstruction, and no therapy is needed. In the other type, the ureter descends normally to approximately the level of L3, where it curves back upward in the shape of a reverse J to pass behind and around the vena cava. Obstruction generally results.

The general symptoms in patients with retrocaval ureter are: flank pain, macrohematuria, recurrent urinal infections or elevation of temperature. Following partial ureteral obstruction, ureteral stones may also occur. In the Slovak literature the treatment of RCU was described in 1986 by Stojkovic and Zvara.



Fig. 2. Spiral C.T. cranio-caudal view.

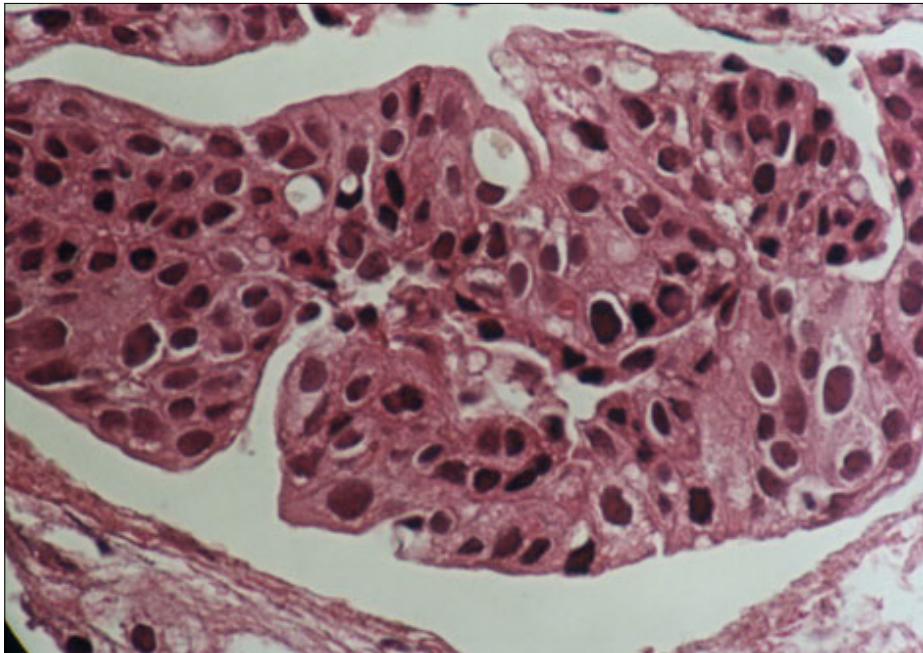


Fig. 3. Histopathology of the suspicious lesion.

In patients without symptoms or urine infection, having either normal flow or a slight obstruction, which is not increasing, periodic observation is recommended.

In symptomatic patients with evident obstruction, decreasing renal function, or continued urinal infection need surgical intervention. The standard surgical intervention is resection of RCU and end-to-end anastomosis or pyelo-ureteral anastomosis per Anderson–Hynes, anterior to the vena cava. This standard surgical intervention can be carried by open or laparoscopic operation. Within the last 6–8 years many surgeons prefer laparoscopic intervention (Ishitoya et al, 1996; Moore et al, 1997; Matsuda et al, 1996). A significant limitation of laparoscopic operation has been a longer operative time compared to open surgery. According to Moore et al (1997) laparoscopic surgery for RCU is successful in 97 %. In our case patient had a RCU with ureteral carcinoma and we were forced to do ureteronephrectomy with a bladder cuff.

In urological anomalies, a quick and correct diagnosis can be difficult. Beside ultrasound examination and urography we want to emphasize the importance of helical C.T. examination, which can be helpful and can reveal other problems. In our case it not only confirmed RCU, but also found a lesion in ureter, which could have been overlooked without helical C.T. In these cases retrograde ureteroscopy can be difficult. Antegrade nephroscopy through the middle calyx, which also allows ureteroscopy, can be very helpful.

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