INTRODUCTION

Malignant lesions of the ureter, also called urothelial tumours are extremely rare, accounting for about 5% to 7% of all renal tumours and about 5% of urothelial tumours. The relatively low frequency of these lesions and the lack of prospective randomized trials do not permit absolute conclusions about the treatment options and their impact on the outcome. The most common presenting symptom of upper tract urothelial tumours is haematuria, this is also true for all renal tumours.

In cases of solitary functioning kidneys, the clinical conditions become even more difficult and the treatment options remain limited, at the cost of chances of tumour recurrence in the due course of time. The outcome associated with the segmental ureterectomy depends largely on the tumour stage and grade. The risk of ipsilateral recurrence of segmental ureterectomy is 44% to 50%, thus, lifelong periodic investigation is warranted because of this risk. Contrary to the segmental ureterectomy, a radical nephroureterectomy in such cases would place the patient on permanent haemodialysis or renal transplant.

CASE REPORT

A 60 years old male presented to the Urology Outdoor Clinic with pain in the left lumbar region for the last 6 months. The pain was mild to moderate in severity, gradual in onset, localized and continuous. There was no history of fever, vomiting, painful micturition or haematuria. He was a known hypertensive for the last 25 years and was on regular medical treatment. He was also a smoker for the last 15 years. On examination, he had blood pressure of 180/110 mmHg and was neither pale nor jaundiced. Abdominal examination as well as the systemic examination was normal.

Complete blood picture revealed haemoglobin level of 14.7 gm/dL and a total leucocyte count of 8.9 x 10^9/L. Urinalysis showed proteinuria and numerous red blood cells per high power field. Serum urea was 36.1 mmol/L (3.3 - 6.7 mmol/L) and serum creatinine 348 umol/L (60 - 110 umol/L). Chest radiograph was normal. Anti-HCV antibodies were positive, however, the liver function tests were within normal range. Ultrasound abdomen was suggestive of left sided hydronephrosis with multiple renal cysts. The right kidney showed irregular margins with a cyst at the upper pole. Plain X-ray KUB did not show any radio-opaque calculus. Intravenous urography, which ideally should not have been performed with such deranged renal profile, was carried out at a peripheral hospital, which revealed left hydronephrosis with multiple renal cysts. The right kidney showed left hydronephrosis with dilated left upper ureter and non-excretory right kidney (Figure 1). So a provisional diagnosis of left ureteric obstruction due to ureteral stricture or radiolucent calculus was made.

Accordingly, ureterorenoscopy (URS) was performed on the left side. This revealed a tight stricture at the middle third of the left ureter which could not be negotiated with the ureterorenoscope or the guide wire. So exploration of the left ureter was contemplated. This showed a dilated upper third and proximal middle third of left ureter. Upon ureterotomy, a papillary growth was seen in the middle third of the left ureter. The growth was excised, sent for histopathology and a double J-stent was inserted.
was placed. The histopathology report was suggestive of transitional cell carcinoma (TCC) of the ureter, so the patient was further investigated. Contrast enhanced computerized tomography of the abdomen and pelvis was performed. Left kidney was larger in size with multiple renal cysts, the largest one (6 x 5 cm) noted at the upper pole. Proximal ureter was dilated and a ureteric stent was seen in situ. A 6 cm segment of soft tissue growth was seen around the mid segment bypassed by the stent. Contrast excretion was also noted. Right kidney was normal in size but the outline was irregular and a 2.5 cm simple renal cyst was also seen. No contrast excretion was noted on the right side (Figure 2).

The patient was diagnosed as having a single functioning left kidney with transitional cell carcinoma of the left ureter. This was a difficult situation and the possibility of auto-transplantation was also kept in mind. Exploratory laparotomy was conducted through a mid line incision. Considering the condition of the left kidney with multiple cysts, the chance of auto-transplantation was rejected. So transuretero-ureterostomy of the proximal left ureter with the right native ureter was performed over a double J-stent. The rest of the left ureter alongwith the tumour and a cuff of bladder (segmental ureterectomy) was removed and sent for histopathology. The margin of the proximal left ureter was subjected to frozen section confirmation and was found to be free of tumour. The left ureter bed was marked with a piece of stent for subsequent radiotherapy (Figure 3). A drain was placed around the ureteric anastomosis and another one in the pelvis, the wound was closed in layers. The postoperative period was quite rewarding. Initially there was some collection in the drain around the anastomosis but this later regressed and the patient made a smooth recovery. On the 6th postoperative day, the serum urea was 11.8 mmol/L and creatinine 205 umol/L. The patient was discharged on the 8th postoperative day after the removal of the drains. The histopathology confirmed a 0.6 x 0.4 x 0.4 cm tumour (residual) of high grade papillary urothelial carcinoma left ureter without muscle invasion (T1 lesion).

He was regularly followed-up and was managed by the oncologist with adjuvant radiotherapy. Three months after the surgery, his serum urea was 5.6 mmol/L and serum creatinine 93 umol/L. On 3, 6, 9, 12, 18 months follow-up he was found asymptomatic. The urine cytology was negative for malignant cells, ultrasound KUB did not show any progressive dilatation / obstruction and cystoscopy was normal on all the occasions. The IVU performed after one year showed a normal upper tract. However, after 18 months of surgery he was lost to follow-up.

**DISCUSSION**

The upper urinary tract tumours can be defined as neoplastic growths that affect the lining of the urinary tract from the calyces to the distal ureter. The frequency of these urothelial tumours is increasing, though, they represent a small percentage of all urothelial neoplasms. Synchronous bilateral urothelial tumours of the upper urinary tract are very rare. Ureteral tumours occur more commonly in the lower than in the upper ureter. About 70% are seen in the distal ureter, 25% in mid ureter and 5% in the proximal ureter. Synchronous bilateral urothelial tumours of the upper urinary tract are very rare. Ureteral tumours occur more commonly in the lower than in the upper ureter. About 70% are seen in the distal ureter, 25% in mid ureter and 5% in the proximal ureter. There is a high rate of recurrence in the distal ureteral stump in patients treated with nephrectomy and incomplete ureterectomy. Conversely, TCC rarely recurs proximal to the level of...
resection of a ureteral lesion. This phenomenon may be a reflection of the downstream implantation. Patients with the upper tract tumours are at risk for development of carcinoma bladder, with an estimated incidence varying from 15% to 75% within 5 years of the development of the upper tract carcinoma.6 This high incidence of metachronous carcinoma bladder suggests that routine bladder surveillance should be performed. On the other hand, in cases of carcinoma bladder, there is a 2 - 4% incidence of upper tract tumours. The upper tract urothelial cancers are often associated with poor prognosis. Nineteen percent (19%) cases present initially with metastatic disease. The ureteral tumours have an even poorer prognosis as compared to the renal pelvic urothelial cancers. TCC accounts for more than 90% of these tumours. Seven percent are squamous cell carcinomas while adenocarcinomas are less than 1%. Rarely, sarcomas may also be seen. The TCC is seen in 58% renal pelvis, 35% in ureter, 7% in both renal pelvis and ureter and bilateral involvement is found in 2 - 5% cases.

Tobacco smoking is the factor most strongly associated with upper tract TCC and increases the risk by more than 3-fold, considered the cause in 70% of upper tract tumours in men and 40% in women. In this case, a 15 years history of smoking seems to be contributory.

Radical nephroureterectomy with the resection of the bladder cuff remains the gold standard for the treatment of upper tract tumours, especially for large, high grade and invasive, and for large multifocal or rapidly recurring, medium grade, non-invasive tumours of the renal pelvis or proximal ureter.7 This is being performed laparoscopically as well. Endoscopic resection or fulguration can be appropriate in selected patients with small low grade lesions. Technological advances have led to an increase in endoscopic management of upper tract TCC. However, all published data are small, retrospective with a short follow-up and high recurrence rate. Endoscopic management of upper tract TCC is particularly suited to small (< 1.5 cm), low grade and non-invasive tumours, in comorbid patients who are at high risk for major radical surgery or who have compromised renal function (including a single functioning kidney), and patients who refuse radical surgery.8 Open segmental ureterectomy (with uretero-ureterostomy, as in this case) is indicated in non-invasive low grade tumours of the proximal ureter or mid ureter that are too large for complete endoscopic ablation, and for high grade or invasive tumours where renal function preservation is the main goal. Segmental ureterectomy can also be performed in distal ureteral tumours, combined with direct neocystostomy or ureteroneocystostomy with a bladder psoas muscle hitch or a Boari flap. The ipsilateral recurrence after conservative treatment of ureteral tumours is more commonly found distal to the lesion; however, proximal ones can also be seen. Thus, there is a lifelong need for a follow-up.

A standard surveillance protocol consists of cystoscopy and selective urine cytology at 3-month intervals postoperatively for the first year and every 6 months during the second year. Excretory urography or retrograde ureteropyelography can be performed at 3- to 6-month intervals to evaluate the upper tract. Ureteroscopy is the most sensitive tool in detecting recurrence and is performed routinely at 3-month intervals initially, with the frequency increasing to 6 months after the first year. At 2 - 5 years, cystoscopy and ureteroscopy are continued at 6-month intervals. Ureteroscopy with biopsy and cytology yields a sensitivity of 93.4% and specificity of 65.2%. In the same series, surveillance with retrograde pyelography had a sensitivity and specificity of 71.7% and 84.7%, respectively.10

REFERENCES