Malignant Glomus Tumor of the Ureter

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ABSTRACT

Although glomus tumour is usually seen in the subungual region of the extremities, it may rarely occur in visceral organs. Approximately 1% of all glomus tumours are malignant. Malignant glomus tumours of the urinary system are extremely rare. This paper presents a case of malignant glomus tumour of the left ureter in a 41-year male patient who had undergone a simple left nephrectomy 4 years ago and presented with recurrent macroscopic haematuria. Uretectomy and mass excision were performed. No progression was observed during the 6-month post-operative follow-up. The tumour can be differentiated from other tumours by pathological and immunohistochemical examination following surgical excision. Because there are very few cases, the treatment protocol is not entirely clear. However, complete removal of the tumour is an effective treatment and can prevent local recurrence.

Key Words: *Malignant glomus tumour, Ureter, Treatment, Urogenital system.*

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INTRODUCTION

Glomus tumour is a mesenchymal tumour composed of modified smooth muscle cells involved in thermoregulation.[1] Although they usually occur in the subungual region in the extremities, they may rarely occur in the visceral organs. There are very few cases reported in the urinary system, and only a few of them are malignant glomus tumours.[2] No case of malignant glomus tumour of the ureter based on literature screening has been reported to date.

CASE REPORT

A 41-year male presented with macroscopic haematuria and abdominal pain for one month. It was learned that the patient had undergone recurrent left ureter endoscopic stone surgery 5 years ago and left simple nephrectomy due to a non-functioning kidney 4 years ago. Atrophy and chronic pyelonephritis were reported in the pathological examination of the kidney. On examination, a mass of approximately 10 cm was palpated in the suprapubic region. İn Magnetic resonance imaging (MRI), ın the middle part of the left ureter, a mass associated with the ureter was detected; it was approximately 97 × 75 mm in size in the axial plane, had heterogeneous intensity at T1AG and T2AG as well as enhancement in post-contrast series with accompanying bleeding areas (Figure 1).

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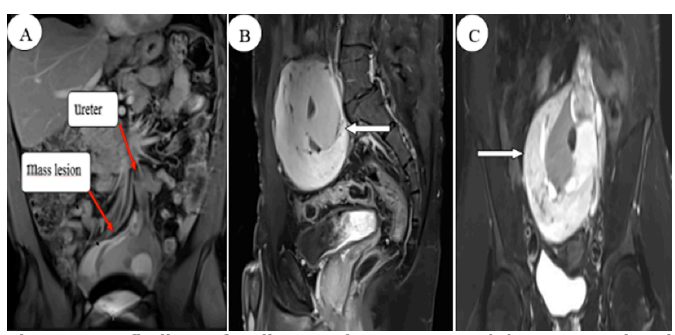

Figure 1: MR findings of malignant glomus tumour. (A) Mass associated with the ureter (T1 image). (B) Image of the mass in the sagittal section (T1 image). (C) Image of the mass in the coronal section (T2 image).

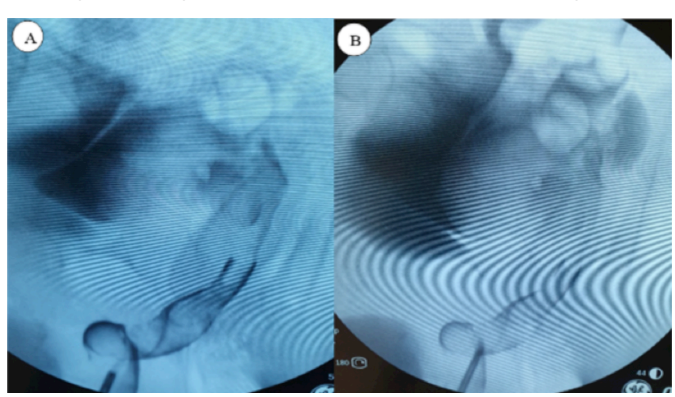

Figure 2: Left retrograde ureterography images. (A, B) Passage of contrast material administered to the ureter to the mass cavity.

Although the clinical presentation was compatible with transitional cell carcinoma (TCC), radiological findings were not compatible with TCC. Left ureterorenoscopy was performed on the patient. Organised hematoma was observed in the ureter lumen, and the ureter was dilated and tortuous. Retrograde ureterography showed that the mass detected on MRI was linked to the

middle part of the ureter and the mass cavity was filled with a contrast agent (Figure 2).

The mass was removed together with the ureter by a sub-umbilical vertical incision. It was observed that the well-circumscribed mass was independent of the surrounding muscle tissues and no invasion was seen anywhere except the ureter. On macroscopic examination, it was noted that the encapsulated mass originated from the ureter and that the mass cavity opened into the ureter lumen (Figure 3).

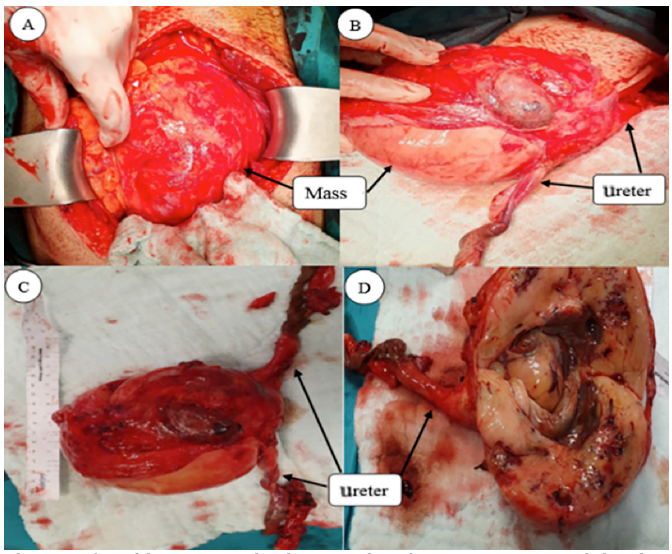

Figure 3: (A, B) intraoperative images; (C, D) 10 × 8 cm mass originating from the left ureter linked to the ureter lumen.

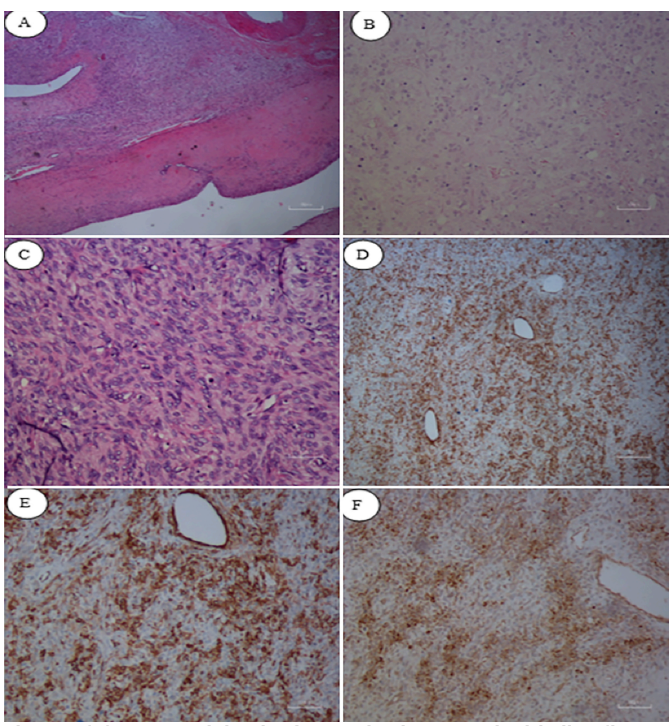

Figure 4: (A) Tumour originating from under the ureteral epithelium (hematoxylin and eosin (H&E), ×40). (B) Tumour necrosis area (H&E 40×). (C) Increased vascularity and mitosis in tumour cells (H&E, ×200×). (D) Smooth muscle actin (SMA) immunohistochemical staining in tumour cells (×40). (E) SMA immunohistochemical staining in tumour cells (×100). (F) Focal desmin immunohistochemical staining in tumour cells (×40).

Pathological and immunohistochemical examination revealed a malignant glomus tumour originating from under the ureteral epithelium. There was no tumour in the surgical margins. Immunohistochemical examination revealed the following: smooth muscle actin (SMA), (+); vimentin, (+); desmin, patchy (+); CD34, (−); and CD31, (−) (Figure 4).

Post-operative positron emission tomography (PET-CT) showed no metastases or residual tumour. No adjuvant therapy was administered to the patient. No local recurrence or metastasis was observed during the post-operative 6-month follow-up.

DISCUSSION

Approximately 1% of glomus tumours are malignant. Malignancy criteria were defined as deeply located tumours, tumours larger than 2 cm, tumours containing atypical mitotic figures or moderate to high nuclear atypia, and tumours containing more than five mitotic figures in 50 high-power fields. The presence of these characteristics has been associated with the risk of metastasis.[3]

Glomus tumours in the urinary system may present with nonspecific findings such as flank pain, abdominal pain, and haematuria.[2] This patient also had recurrent macroscopic haematuria and abdominal pain. Because glomus tumours in the urinary system do not have characteristic radiological findings, it is challenging to establish a preoperative diagnosis. For the diagnosis, pathological examination and immunohistochemical evaluation are required after surgery or biopsy.[2]

In immunohistochemical studies, glomus cells show positive immunoreactivity with SMA and vimentin, whereas vascular endothelial markers including CD31 or CD34 were negative. But, in one published report, cells were positive for CD34 in about 41.7% of the cases. However, it was reported that desmin positivity may vary.[4]

Local recurrence rate following surgery in benign glomus tumours has been reported to be 10% and is generally attributed to inadequate excision.[3] Malignant glomus tumours are very aggressive. It has been reported that most patients diagnosed with malignant glomus tumours in visceral organs die due to tumour progression immediately after diagnosis.[1] In the study performed by Folpe *et al*., the metastasis rate was 38% in dermal malignant glomus tumours. Lamba *et al*. reported that in the malignant glomus tumour of the kidney, the patient's condition progressed and he died 6 months after diagnosis despite palliative chemotherapy.[5] In the malignant glomus tumour of the bladder reported by Shim *et al*., peritoneal invasion and lung metastases were detected. Despite chemotherapy, the patient died 2 months after diagnosis.[6] In the malignant glomus tumour of the kidney reported by Chen *et al*., no progression was observed in the 6-month follow-up.[2] In the present patient, no progression was observed in the first 6 months of follow-up.

Malignant glomus tumours of the urinary system are extremely rare. These can be differentiated from other tumours by patho-

logical and immunohistochemical examination following surgical excision. Because there are very few cases, the treatment protocol is not entirely clear. However, complete removal of the tumour is an effective treatment and can prevent local recurrence.

PATIENT'S CONSENT:
Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
MD: Design, analysis, and drafting.
AT: Data collection.
IY: Data analysis and manuscript review.
AA: Assisted in drafting the data.

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