

## Case Report

# Primary extraskkeletal osteosarcoma of the ureter: report of a case

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**Abstract:** Extraskkeletal osteosarcoma (EEO) is a rare malignant mesenchymal tumor that grows in soft tissue without attaching to skeleton. Extraskkeletal osteosarcoma arising in the ureter is extremely rare and only 3 cases have been reported in detail to date. In our study, an 81-year-old man presented with a 1-month history of painless naked hematuria. Computed tomography (CT) scan revealed that there was a solid tumor in the left distal ureter. Subsequently, radical resection was performed. Immunohistochemical staining of specimens showed that the tumor cells were positive for vimentin and Ki-67, but negative for other specific markers. The findings favored the primary ureteral osteosarcoma. Adjuvant radiotherapy was recommended but was refused. Four months later, regular clinical follow-up revealed no evidence of recurrence. The aim of present study is to report an additional case of ureteral osteosarcoma, focusing on the characteristics, including pathogenesis, diagnosis, treatment and outcomes.

**Keywords:** Extraskkeletal osteosarcoma, ureter, characteristics, outcomes, case report

### Introduction

Extraskkeletal osteosarcoma (EEO) is a rare malignant tumor arising from soft tissue, accounting for approximately 4% of all osteosarcoma [1]. The tumor is likely to occur in middle-aged and elderly patients, and mainly located in the deep soft tissue of the retroperitoneum, chest, breast and neck, but rarely originating from internal organs [2]. Osteosarcoma primarily originating from the ureter is specifically sparse, and only 3 cases had been reported in the literature [3-5]. In this study, we portrayed an 81-year-old man, who presented with left ureteral mass discovered by CT-scan after 1-month history of painless naked hematuria.

### Case report

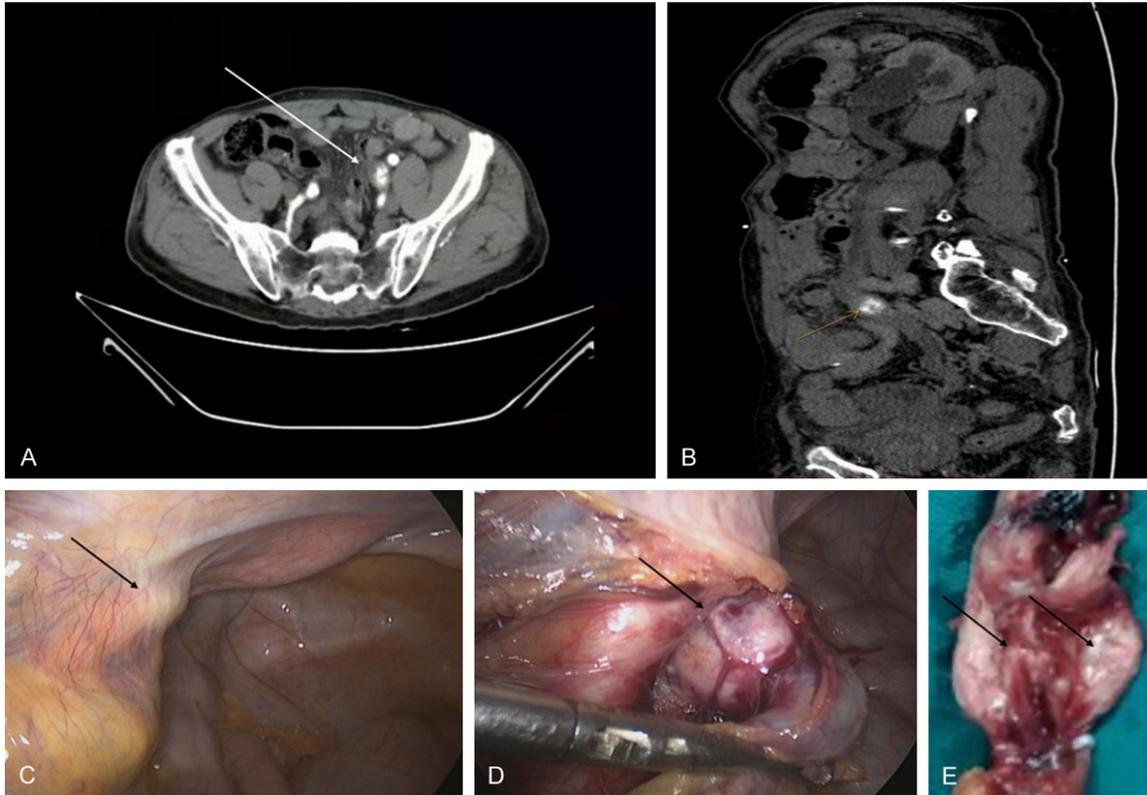
In September 2018, an 81-year-old man was admitted with no previous history of cancer who suffered from painless naked hematuria with passage of clots for about 1 month. He had a history of hypertension. Beyond percussion pain in the region of the left kidney and left ureter, the general physical examination was normal. No abdominal mass on palpation was

found. Urine cytology examination was negative in finding the malignant cells. Spiral computed tomography (CT) scan of urinary system revealed a solid mass with calcification in the left distal ureter accompanied by upper ureter hydronephrosis and kidney hydronephrosis (**Figure 1A** and **1B**). Ureteroscopy was subsequently performed, but failed, because the ureter was filled with blood clots and necrotic tissues. No secondary lesions were found in chest CT-scan.

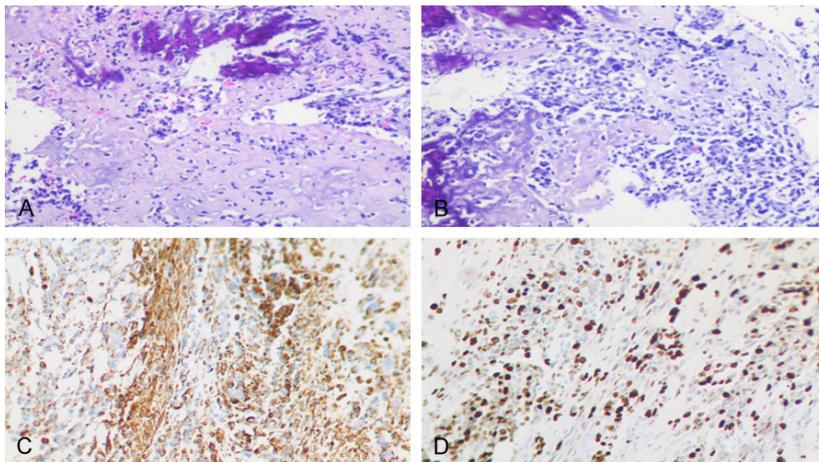
After ruling out surgery contraindications, uneventful laparoscopic radical nephroureterectomy on the left side was performed with a preoperative diagnosis of left ureteral tumor, hematuria and hypertension.

During the operation, the tumor appeared about 2 cm in diameter, round in shape and hard in texture, located in the ureter at the level of the crossing of the iliac vessel (**Figure 1C** and **1D**). It had no obvious adhesion with surrounding tissues. To the naked eye, the tumor was grey, about 2 cm × 1.5 cm × 2 cm in size, located at about 5 cm from the distal end of the ureter. The cut surface of the tumor was grayish white, with a feeling of gravel and calci-

## Osteosarcoma of the ureter



**Figure 1.** A and B. CT-scan revealed a solid mass with calcification in the left distal ureter. C and D. A solid tumor, in the ureter at the level of the crossing of the iliac vessel. E. The cut surface of the tumor was grayish white with hard texture and appeared to invade the muscular layer with the naked eye.



**Figure 2.** (A and B) Observation under HE staining microscope revealed focal necrosis and focal area containing osteoid. Immunohistochemical staining showed that the tumor cells were strongly positive for vimentin (C), moderately positive for Ki-67 (D).

The tumor was limited to the ureter and the resection margin was negative. No tumor cells were detected in the resected end of ureter or kidney. Immunohistochemical staining of specimens showed that the tumor cells were positive for the expression of vimentin and Ki-67 (**Figure 2C** and **2D**), but negative for CK, LCA, GATA3, CK5/6, P63, Uroplakin-III and Pax-8, supporting the diagnose of extraskeletal osteosarcoma of the ureter.

fication in parts of the area, and appeared to invade the muscular layer (**Figure 1E**). Histopathology subsequently revealed focal necrosis and focal area containing osteoid (**Figure 2A** and **2B**), invading the entire ureteral wall.

About 1 week later, the patient recovered uneventfully from surgery and was discharged. Adjuvant radiotherapy was recommended but was refused. Four months later, the patient was doing well, and regular clinical follow-up

revealed no evidence of recurrence, including local or metastatic lesions.

### Discussion

Extraskelatal osteosarcoma (EEO) is a kind of malignant tumor which grows in soft tissue and has the ability to form osteoid matrix, bone tissue and chondroid substance [1]. So far as is known, there are three types of pathogenesis. Firstly, during embryonic development, the mesoderm remains, and then bone forms, resulting in the proliferation of osteogenic elements. Secondly, any part of connective tissue that may be stimulated by some external or internal stimulus, such as trauma, bleeding, inflammation, etc., has local biochemical changes. The stimulated fibroblasts can be metaplastic, forming osteoblasts and developing into osteosarcoma. Thirdly, ectopic osteochondral can be transformed into osteosarcoma [6].

EEO has no characteristic changes in clinical manifestation and imaging examination. It has some differences with osteosarcoma in clinical features as follows. Firstly, EEO is not as sensitive to chemotherapy as osteosarcoma. Secondly, the 5-year survival rate of EEO is 25%-37%, while osteosarcoma is about 60%. Thirdly, EEO is more common in middle aged and elderly people, while osteosarcoma is more common in adolescents [7]. The diagnosis of EEO is mainly based on the pathological examination, and should be distinguished from carcinosarcoma, transitional cell carcinoma with interstitial ossification, and transitional cell carcinoma with pseudosarcoma.

Extraskelatal osteosarcoma of the ureter (EOOU) is a very rare soft tissue tumor with high-malignancy. So far, only 3 well-documented cases have been reported in the literature. The tumor is solid, polypoid, and gravelly on the cut surface. Furthermore, bleeding and necrosis may be found in some part of the tumor. The most common symptoms of EOOU are hematuria, dysuria, frequent urination and recurrent urinary tract infection [3-5]. Radical surgical resection is the main treatment of choice for EOOU. In 3 previous cases of EOOU, 1 case presented with symptomatic hydronephrosis, 1 case presented with flank pain and anuria, and 1 case presented with hematuria, urination pain and dysuria. Similar to our current case, all of the 3 cases were performed with surgical

resection, and all of the 3 cases accepted no adjuvant chemotherapy or radiotherapy.

This case was also found by the symptom of naked hematuria. Adjuvant therapy, such as chemotherapy or radiotherapy, is helpful for improving the survival rate and reducing recurrence and transfer rates in patients with EEO [8]. Considering the patients' age, radiotherapy was recommended without chemotherapy but was refused.

As for the outcomes, EEO has a poor prognosis, due to its strong invasiveness. Distant metastasis may occur in advanced stages of EEO, and the most important location of metastasis is the lung [1]. In 3 previous cases of EOOU, 1 case developed tumor recurrence 3 months after surgery, and 2 cases exhibited no recurrence up to 6 months after surgery. In this case, the tumor was discovered relatively early, and no significant metastasis had occurred to date. We will pay close attention to the long outcome of this patient.

### Acknowledgements

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### Disclosure of conflict of interest

None.

### Abbreviations

EEO, extraskelatal osteosarcoma; EOOU, extraskelatal osteosarcoma of the ureter.

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