First case of a primary osteosarcoma of the ureter: diagnostic findings, course of disease and treatment

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Case report

In October 2005, a 61-year-old patient was admitted with symptomatic right sided hydronephrosis. She was treated for breast cancer in 2003 with radical mastectomy and chemotherapy and for cervical carcinoma in 1983, which was cured by a Wertheim-Meigs operation and radiotherapy.

An intravenous pyelogram was performed, which demonstrated occlusion of the right distal ureter (Figure 1). A subsequent ureteroscopy revealed a hard, round mass in the distal ureter that was not removable and occluded the lumen. Biopsies were taken and histopathology revealed a highly pleomorphic neoplasm composed of myxoid spindle cell sarcomatous elements and more closely packed spindle cells with malignant osteoid production. Distal ureteric resection with bladder-cuff and ureterocystoneostomy were performed (R0-resection). The patient made a good recovery from surgery and was discharged. Adjuvant chemotherapy was recommended but rejected by the patient.

In January 2006, the patient returned to the outpatient clinic, suffering from macrohaematuria and right sided flank pain. Cystoscopy suggested tumour recurrence in the region of the neo-ostium. Biopsies were taken and histopathology revealed an early relapse of the previous osteosarcoma. To define the tumour stage, a CT-scan was performed and revealed a dilated right renal pelvis (Figure 2) and a grossly dilated right ureter without further evidence of extraluminal tumour relapse or evidence of metastatic disease. An ureteronephrectomy with partial bladder resection was conducted. Histology confirmed local and extraureteral tumour relapse (R2-resection). Furthermore, a metastasis in the renal pelvis, infiltrating the renal parenchyma, was detected. Palliative chemotherapy, according to the COSS (Co-operative osteosarcoma study group) trial,¹ was initiated.

In July 2006, the patient was seen in the outpatient clinic because of the presence of white pieces of tissue in the urine. Imaging (Figure 3) and cystoscopy revealed significant massive tumour growth that was confirmed by laparotomy. Histology showed metastatic lesions in the abdominal wall, the peritoneum, the ileum and the sigmoid colon. The patient received palliative care but died 11 months after initial presentation.

Discussion

We believe this to be the first reported case of primary ureteral osteosarcoma. Previously, rare cases of primary osteosarcoma of the bladder² and the kidney³ have been described.

Extraosseous osteosarcoma are known to have a poor prognosis despite radical surgical resection and chemotherapy.⁴ In 2005, Goldstein-Jackson et al.⁵ reported a more favourable prognosis in extraskeletal osteosarcoma when treated as conventional osteosarcoma according to the COSS protocols.
In their retrospective study, they described a heterogeneous group of 17 patients with extraskeletal osteosarcoma, most notably located in the lower extremity (none in the urogenital system), and found an estimated 5 year overall survival of 77% and an estimated 5 year event-free survival of 56% after surgical resection and adjuvant chemotherapy.

Radiation therapy is known to be associated with subsequent development of bone and soft tissue sarcomas, including extrasosseous osteosarcoma. The time interval between radiotherapy and development of sarcoma can be prolonged, with a mean interval of \(~12\) years. Our patient underwent radiation therapy due to a cervix carcinoma 22 years before the occurrence of the extrasosseous osteosarcoma. Unusual calcifications (Figure 3) may be suggestive of this neoplasm. Nevertheless, extraskeletal osteosarcoma should be distinguished from other tumours that may be associated with bone formation, such as carcinosarcomas and transitional cell carcinomas with osseous metaplasia of their stroma, both of which have a better prognosis than osteosarcoma.
In osteosarcoma of the urogenital system, diagnosis is mostly made in advanced stages of disease because of weight loss, flank pain or macrohaematuria. In this case, the tumour presented relatively early, imitating a urolithiasis with hydronephrosis and flank pain. Despite initially successful radical surgery, the patient died <1 year after initial treatment demonstrating the highly malignant characteristics of this tumour entity and the need for immediate chemotherapy in addition to surgery.

References