

# Sarcoma of the bladder with metastasis to the left ventricle

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## Abstract

Genitourinary sarcomas are rare clinical entities. Current information on these tumours is sparse and anecdotal. Cardiac metastasis from genitourinary tumours in general is an exceedingly rare phenomenon and has previously been reported only in relation to carcinomas. We report the first case of sarcoma of the bladder with metastasis to the heart causing sudden death.

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## Introduction

Genitourinary sarcomas are rare malignant tumours of mesodermally derived extraosseous tissues composing only 1%–2% of all genitourinary tumours.<sup>1,2</sup> Current information on these tumours is sparse and is based on anecdotal reports and small case series. We report the first case of sarcoma of the bladder with metastasis to the left ventricle of the heart.

## Case report

A 77-year-old female patient presented to our emergency department with a history of frank hematuria. She had been treated for recurrent urinary tract infections in the preceding 3 months. Dipstick examination of the urine then revealed microscopic hematuria with normal hematological and biochemical blood investigations. Her past medical history included 3 myocardial infarctions, an abdominal aortic aneurysm repair, a carotid endarterectomy and peripheral vascular disease. Ultrasound of her bladder revealed a 10 × 11 × 12-cm intravesical mass with a mixed echo texture, raising the possibility of a clot or tumour. The patient's ureters and kidneys were reported normal. A preoperative echocardiogram, performed in light of her previous cardiac history and extensive vascular disease, showed a normal-sized ventricle with normal function.

The patient underwent a cystoscopy that showed a large whitish mass almost completely occluding the bladder lumen. A partial transurethral resection of bladder tumour (TURBT) was performed and tissue was sent for histological examination. Initial histology showed infarcted material with focal neutrophilic infiltrate in some areas. There was no viable tumour tissue and a repeat biopsy was advised. Subsequent CT scan of the abdomen and pelvis showed a large residual mass in the right lateral wall of the bladder with areas suggestive of bladder wall invasion. There was no evidence of pelvic or abdominal metastatic spread.

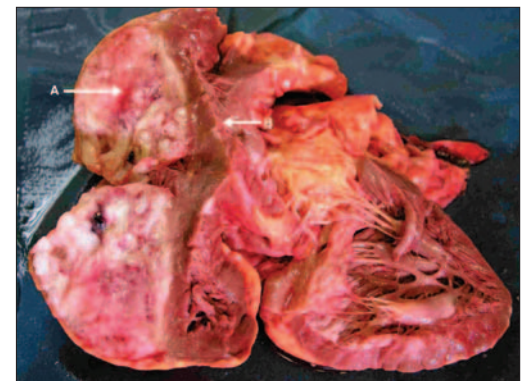
A repeat TURBT with piecemeal removal of the bladder tumour was performed to obtain a diagnosis, and a catheter was left in situ to overcome

major voiding problems. Histology showed a high-grade undifferentiated sarcoma with some features to suggest neural differentiation. An immunohistochemical analysis of the tumour tissue ruled out carcinoma and showed positive staining for some neural markers, specifically neuron-specific enolase and PGP 9.5. Markers for smooth muscle differentiation (leiomyosarcoma) were negative.

Isotope bone scan did not reveal any bony metastasis. The patient was optimized to undergo a radical cystectomy 5 weeks after her initial presentation but unfortunately died suddenly 6 days before the operation. Postmortem examination revealed hemorrhagic pericarditis and a nodular tumour arising from the interventricular septum extending toward the pulmonary valve outlet (Fig. 1). Histological examination of this metastatic lesion was identical to that found in the bladder tumour. The cause of death was accordingly reported as hemorrhagic pericarditis secondary to intracardiac metastasis from a bladder sarcoma.

## Discussion

Genitourinary sarcomas are rare tumours that account for 1%–2% of all genitourinary



**Fig. 1.** Metastatic tumour (A) obstructing the left ventricular outflow tract. B = the septum.

tumours and less than 5% of all sarcomas.<sup>1,2</sup> They occur at extremes of life,<sup>1</sup> with rhabdomyosarcomas being the more common variety in children and leiomyomas more common in adults.<sup>1,2</sup> There is a range of histological types in between, including fibrosarcoma, angiosarcoma and extrasosseous osteosarcoma.<sup>1</sup>

Painless frank hematuria is the most common presenting symptom. Urinary frequency, dysuria, nocturia, urgency, urinary retention and recurrent urinary tract infections are less common presentations.<sup>1-4</sup>

Suggested contributory factors to the development of these tumours include genetic links, embryological mesenchymal remnants, radiation and chemotherapy.<sup>1</sup> However, no definite cause-effect relationship has been identified.

Considering the rarity of these tumours, there is no single accepted staging system. However, a system developed at the Sloan Kettering Institute constituting tumour grade, size (< or > 5 cm), depth of invasion, presence of metastatic disease and retinoblastoma gene product has proved useful in terms of prognosis and predicting survival rates.<sup>2</sup>

Recommendations for treatment vary and reports are based on small studies owing to the rarity of this tumour. In some centres radical cystectomy is recommended for localized disease,<sup>5</sup> while in others partial cystectomy is used provided adequate surgical margins can be obtained.<sup>6,7</sup>

The role of radiotherapy is unclear, with some centres using it for documented or presumed microscopic disease and in larger bulkier tumours if surgical margins are positive after resection.<sup>6,7</sup>

Preoperative cytoreductive chemotherapy has been used with cisplatin and doxorubicin, together with postoperative adjunctive chemotherapy in sensitive and bulky tumours.<sup>6</sup> Advanced and metastatic disease is treated with chemotherapy, doxorubicin and ifosfamide being the most active agents.<sup>2</sup>

Metastasis to the heart from bladder tumours is an exceedingly rare phenomenon with only 5 reported cases in the English literature, all from transitional cell carcinoma histology.<sup>8-12</sup> We now report the first case of bladder sarcoma with metastasis to the heart and propose that this phenomenon may be one of the less commonly recognized causes of sudden death in patients with bladder sarcomas and that imaging of the heart may be indicated as part of the workup for patients with advanced bladder sarcomas. It is surprising that the

metastatic lesion in the heart did not show up on the preoperative echocardiogram in our case. However, the echocardiogram was performed as part of the preoperative workup of an elderly patient with an extensive previous cardiac history and it is possible that this lesion may have been missed because we were not looking for it. Another possibility is that the sarcoma may have experienced explosive growth during the time between the echocardiogram and the patient's death and that it may have initially been too small to see on the echocardiogram. Nevertheless, a high index of suspicion is needed when evaluating the hearts of patients with advanced bladder sarcomas.

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