Renal sarcoma and associated malignant pulmonary embolism: a report of 2 cases

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Abstract

Renal sarcoma with venous tumour thrombus is usually an aggressive malignancy that necessitates complete surgical extirpation to achieve cure. Due to the rarity of these tumours, clinicians rely on case reports to better understand and treat patients with this disease. We recently encountered 2 patients with renal sarcoma who developed malignant pulmonary embolus. Our cases, combined with those previously published, suggest renal sarcoma tumour thrombus is at high risk for spontaneous and intraoperative embolization. This report details our experience and outlines measures that may decrease the rate of venous tumour embolization in patients with sarcoma.

Case Presentation and Management

Case 1

A 17-year-old man presented with minor flank trauma and gross haematuria. Magnetic resonance imaging revealed a large right renal mass and associated IVC tumour thrombus (Fig. 1). Renal biopsy suggested high-grade sarcoma. Metastatic workup was negative. The case was discussed at the Children’s Hospital of Eastern Ontario pediatric oncology rounds, and it was decided that the best course of management was surgical resection without neo-adjuvant radiotherapy or systemic chemotherapy. Right nephrectomy, thrombectomy and extensive lymph node dissection were performed without apparent complication. Postoperatively, the patient was dyspneic and CT scan of his thorax revealed bilateral PE (Fig. 2). Doppler imaging of his IVC and leg veins revealed no source of bland thrombus, which is highly suggestive of pre- or intraoperative malignant PE. The patients’ histopathology was consistent with a high-grade undifferentiated sarcoma with focal rhabdoid differentiation. Immunostaining revealed focal reactivity for AE1 and AE3, vimentin and CD99. Negative reactivity was seen for S100, muscle specific actin, chromogranin and leukocyte common antigen. The patient died 3 months after the surgery. He had diffuse metastatic disease to bone and lung.

Case 2

A 43-year-old woman presented with dyspnea, hematuria and left flank pain. She had been diagnosed and treated with anticoagulation for a presumed benign PE. Chest and abdominal CT scans revealed a large left renal mass with associated IVC extension (Fig. 3) and bilateral proximal PE (Fig. 4). Pulmonary endarterectomy could not completely remove the malignant emboli, because they had invaded the bronchial walls. Nephrectomy with thrombectomy were undertaken for palliation of local symptoms and Budd-Chiari syndrome. The thrombus invaded the IVC wall and removal was incomplete. Histopathology of both the pulmonary emboli and the kidney tumour were consistent with clear cell sarcoma. Immunostaining was reactive for vimentin and negative for all cytokeratins.
S100, HMB45 and epithelial membrane antigen. The patient died 1 month after surgery.

**Discussion**

This is the first report to highlight the association of PE with advanced renal sarcoma. Currently, of 12 sarcoma cases with IVC thrombus, 5 (42%) had malignant PE. This prevalence may be artificially elevated due to publication bias, since unusual or unfortunate events are more frequently reported. Nevertheless, the increased incidence of emboli suggest sarcoma tumours are more susceptible to fragmentation than renal cell carcinoma.11

Unfortunately, in the absence of tumour histology, it is not possible to differentiate renal sarcomas from renal cell carcinomas.12 Although certain properties, such as young patient age, tumour origin from the renal capsule or renal sinus, decreased tumour vascularity, rapid tumour growth and absence of lymphadenopathy associated with a large primary tumour favour sarcoma, these findings can also be associated with renal cell carcinoma. Therefore, we suggest treating all patients with these characteristics as high risk for malignant PE.

Currently, surgical resection is the primary treatment of organ confined and locally advanced renal sarcoma. However, pediatric patients and patients with unresectable tumours may benefit from radiotherapy and chemotherapy. It is therefore recommended that a multidisciplinary oncology group vet these cases before surgery.

In patients with confirmed or suspected renal sarcoma and venous tumour thrombus, the following recommendations may reduce the risk of malignant embolization:

1) expedient nephrectomy;
2) consideration to preoperative renal angioinfarction to cause thrombus shrinkage; and
3) careful intraoperative thrombus handling with early proximal venous control.

**References**


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