

Oncology

Malignant solitary fibrous tumour of the kidney with an extensive thrombus: A case report and review of the literature



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ABSTRACT

Solitary fibrous tumours (SFTs) are neoplasms originating from mesenchymal cells, usually found in lung pleura. Extraleuritic lesions are extremely rare, with about 60 cases of SFTs of the kidney available in the English literature. We report here the first case of a malignant SFT of the kidney presenting with an extensive vascular thrombus extending to the right atrium with associated pulmonary emboli. We describe management, pathological assessment, as well as radiological and clinical evolution. Our report provides a first therapeutic approach to a critical presentation of a rare pathology, which should help guide management of such disease in future cases.

Introduction

Solitary fibrous tumours (SFTs) are spindle cell tumours arising from mesenchymal cells, usually found in lung pleura.¹ Extraleuritic sites of SFTs such as bone, endocrine and exocrine glandular tissue, and the genitourinary system, have been described. The first case of a SFT of the kidney was reported in 1996,² and 64 reports are currently available in the English literature. Herein, we report the case of a patient who presented to our institution with a malignant SFT of the kidney associated with an extensive venous thrombus extending to the right atrium.

Case presentation

We report the case of a 72 year-old male who presented to the emergency room with a 4-day history of respiratory symptoms. He denied any symptoms of heart failure, hematuria, or lower urinary tract symptoms. A contrast-enhanced thoracic computed tomography (CT) scan revealed multiple pulmonary emboli, as well as a partially-imaged mass at the upper pole of the right kidney. A dedicated abdominal scan revealed a contrast-enhancing, slightly heterogeneous, 6.0 × 4.7 × 5.0 cm endophytic mass of the right renal upper pole with an associated

heterogeneously-enhancing venous thrombus extending into the right atrium, causing a dilatation of the intra-hepatic vena cava up to 4.5 cm. (Fig. 1) There was no evidence of any metastases or lymphadenopathies.

It was decided that the patient would undergo open right nephrectomy, right adrenalectomy, thrombectomy, and inferior cavectomy. After extensive dissection, the inferior vena cava (IVC) was clamped just inferiorly to the right renal vein, but above the left renal vein. This occlusion caused no hemodynamic alteration, suggesting complete IVC occlusion and extensive collateral neovascularization. The patient was then put on cardiopulmonary bypass, with a 7-min hypothermic cardiac arrest allowing for tumour resection. The right atrium was opened, and the thrombus was pushed down and completely extracted through the IVC. Inspection of the IVC revealed probable invasion, and cavectomy was therefore performed on its portion between the right renal vein and the hepatic vein. The surgery lasted just over 8 hours.

The surgical specimen showed a 6.0 × 4.5 × 4.0 cm, beige, homogenous mass involving the renal hilum, renal vein, and IVC. The tumour also invaded the renal capsule and adjacent renal parenchyma superficially, with overall negative surgical margins. Microscopy revealed fusiform spindle cells with collagen bands in patternless arrangements, with occasional areas of hemangiopericytomatous appearance (Fig. 2).

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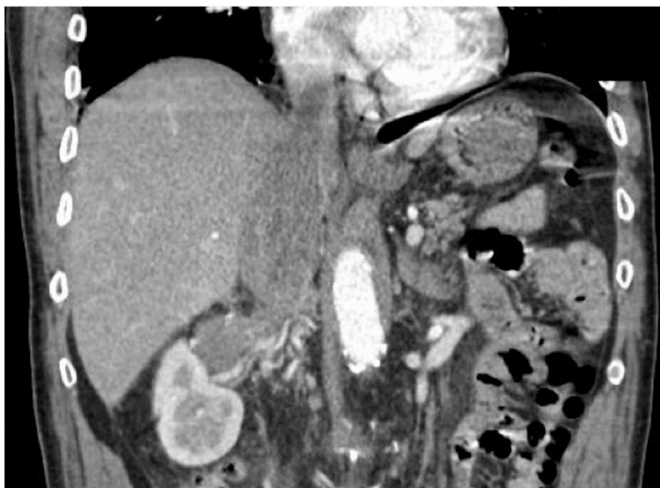


Fig. 1. A dedicated abdominal scan revealed a contrast-enhancing, slightly heterogenous, $6.0 \times 4.7 \times 5.0$ cm endophytic mass of the right renal upper pole with an associated heterogeneously enhancing venous thrombus extending into the right atrium, causing a dilatation of the intra-hepatic vena cava up to 4.5 cm.

The tumour was found to be a malignant form of SFT of the kidney, staining for CD99, CD34 and Vimentin, while being negative for usual urothelial and renal markers. Regions of tumour necrosis, high mitotic activity (> 4 mitoses/10 HPF), and vascular thrombus were all suggestive of malignancy.

The patient was asymptomatic and doing clinically well on follow-up visits up to 9 months after surgery. However, a CT scan performed at that time revealed the growth of multiple bilateral pulmonary nodules, suggesting metastatic progression. Abdominal imaging did not reveal any regional recurrence. Based on newly available data,³ the patient started a treatment protocol of tyrosine kinase inhibitor pazopanib, which he is currently receiving with good tolerance.

Discussion

SFTs are neoplasms characterized by the proliferation of spindle cells originating from fibroblasts and mesenchymal cells, and are

usually found in lung pleura. Other sites include bone, endocrine and exocrine glandular tissue, and the genitourinary system, with kidney being one of the rarest locations. On pathology, tumours are usually tan-gray, homogenous, lobulated, and well-circumscribed, and most often involve the renal capsule.¹ Microscopically, fusiform spindle cells are surrounded by thick, keloid-like collagen bands with patternless arrangements. Immunohistochemical testing usually reveals positivity for vimentin, CD34, and CD99, as well as bcl-2 and epithelial membrane antigen in about a third of cases.^{1,4} On imaging, enhanced CT scan usually reveals well-circumscribed lesions with homogenous enhancement, although zones of heterogenous enhancement can appear with tumour hemorrhage or necrosis. Magnetic resonance imaging shows predominant low-to-intermediate signal density on T1- and T2-weighted-images secondary to high fibrous collagenous content.⁴ The radiological differential diagnosis is long, and immunohistochemical testing usually helps making the diagnosis.¹ While about 90% of SFTs of the kidney are benign, microscopic characteristics such as increased cellularity, pleomorphism, and increased mitotic activity are diagnostic for malignancy.⁴ Tumour characteristics such as necrosis, hemorrhage, size, and vascular thrombus are also suggestive of malignancy.¹

SFTs of the kidney have only been reported in 64 fully-described cases in the English literature. Tumours were usually found incidentally on imaging.⁵ Radical nephrectomy has been the mainstay treatment in most reported cases, with a few patients being managed with partial nephrectomy with good oncological outcomes. Only two other cases in the literature have reported some component of vascular thrombus, both proximal to the renal vein ostium. Simple thrombectomy was performed in both cases.

Given their unpredictable malignant potential, SFTs are considered “intermediate malignant, rarely metastasizing” neoplasms, and all patients should be on long-term follow-up.⁵ Systemic chemotherapy, immunotherapy, and radiotherapy may have a role in the treatment of malignant or metastatic disease, but there are currently no standard protocols or regimen.⁵ A recent multicentre, phase 2 trial showed interesting results for patients treated with pazopanib.³ This study prompted our team to start treatment with this tyrosine kinase inhibitor given the progression of metastatic pulmonary lesions.

Our case is therefore unique in multiple ways. The fact that such an extensive mass presented only with respiratory symptoms, and no hematuria or abdominal pain, is surprising. Also, the extent of the associated vascular thrombus, and the surgery required for resection is a

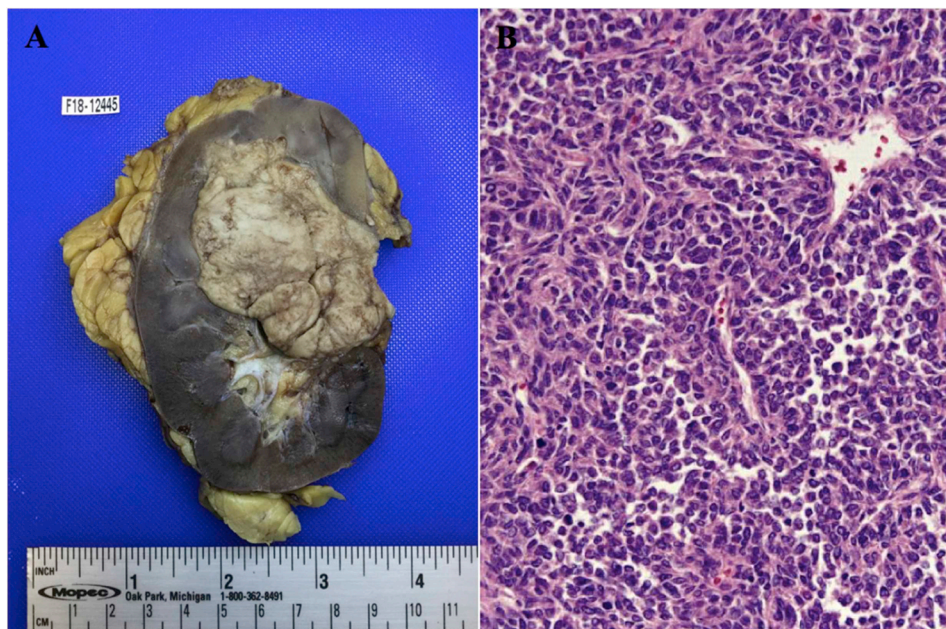


Fig. 2. (A) The surgical specimen showed a $6.0 \times 4.5 \times 4.0$ cm, beige, homogenous mass involving the renal hilum, renal vein, and IVC. The tumour also invaded the renal capsule and adjacent renal parenchyma superficially. (B) Microscopy revealed fusiform spindle cells in patternless arrangements, with occasional areas of hemangiopericytomatous appearance.

first in the literature for this type of tumour. The malignant nature of the renal SFT on histology is also a rarity in itself, as well as its pulmonary metastatic progression. The patient will complete immunotherapy at our institution, and clinical response will be assessed with regular imaging and close follow-up.

Conclusions

We reported what is, to our knowledge, the first case of a malignant SFT of the kidney presenting with a venous thrombus extending into the right atrium. We described clinical and radiological presentation, surgical resection of the tumour, clinical evolution with metastatic progression, and oncological management. We also discussed the state of the literature on SFTs of the kidney. Our report should help guide management of such disease in future cases, and contributes to the literature on the poorly understood clinical course of this entity. This case highlights the importance of including SFTs in the differential diagnosis of a renal mass presenting with a venous thrombus.

Conflicts of interest

The authors have no conflict of interest to declare.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.eucr.2019.100974>.

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