## Case Report

# An Incidental Solitary Fibrous Tumour in the Retroperitoneum, Coexisting with Ipsilateral Atrophic Kidney

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## Abstract

Solitary fibrous tumour (SFT) is a relatively uncommon spindle-cell neoplasm that most commonly arises in the pleura, but which may also arise from other serosal surfaces outside the pleura. However, SFT is now known to affect various serosal surfaces including pericardium, peritoneum, retroperitoneum nasal and paranasal sinuses, thyroid, cavernous sinus or pituitary fossa. The histologic features of this lesion may create diagnostic confusion with a variety of other spindle-cell tumours. To the best our knowledge, no cases with SFT have been previously noted in the retroperitoneum coexisting with atrophic kidney. Herein, we report the unique association of a solitary fibrous tumour in the retroperitoneum coexisting with ipsilateral atrophic kidney in a 60-year-old man and define histopatological findings of this tumour.

**Keywords:** Atrophic kidney, retroperitoneal mass, solitary fibrous tumour, mesothelioma.

## Introduction

Solitary fibrous tumour (SFT) is a rare neoplasm, which was originally termed localized fibrous mesothelioma in the pleura. Later, it was known as solitary fibrous tumour.[1] Solitary fibrous tumours (SFTs) are spindle cell neoplasms, frequently arising in the pleural cavity but they have been described in other serosal surfaces and in nearly every organ such as the pericardium, peritoneum and liver. Importantly, they may occur without an association to a serosal surface as in the mediastinum, orbit, thyroid and nasal cavity.[2,3] The etiology of SFT is unknown and usually recognized in patients between the third or fourth decades of life.[3] On ultrastructural examination, tumour cells show fibroblast-like rather than mesothelial-like features. Immunohistochemically, most of the tumour cells stain strongly for CD34, but do not stain with keratin, desmin, S-100 protein or alpha smooth muscle actin. The unifying characteristic is the positivity of the spindle cells for CD34.[4] The clinical behaviour of SFT is usually benign, but the existence of aggressive cases has been reported both in the pleura and in extrapleural sites.[3,4] A careful immunohistochemical examination is necessary to differentiate SFT from other spindle cell neoplasms with a more aggressive nature.[5] The retroperitoneum is an extremely rare site of origin for SFT. A total of 25 cases of only retroperitoneal SFT, have been documented in recent

literature.[6] However, no cases with SFT have been previously noted in the retroperitoneum coexisting with atrophic kidney. In the present case, we report a solitary fibrous tumour of the retroperitoneum coexisting with ipsilateral atrophic kidney in a 60-year-old man.

## Case Report

A 60-year old man presented with right flank pain for two days in our emergency department (ED). Mild hypertension with a blood pressure of 150/100 mmHg was noted on physical examination. While routine biochemistry and chest radiograph was normal. Renal ultrasonography revealed right atrophic kidney (4x5x6 cm in diameter). Intravenous urography and renal scintigraphy confirmed a right sided nonfunctional kidney, but no additional pathology in the contralateral kidney. The decision for surgery was made on the basis of hypertension and a non-functional kidney. An open simple nephrectomy was performed with a flank incision, because the laparoscopic equipments were not available in our clinic. While dissecting the right kidney and the ureter, a capsulated round mass, 3cm in diameter, lying at the anterior side of the psoas muscle was incidentally detected. This was located 3 cm caudally to the atrophic kidney. The mass was completely excised along with the right kidney. Pathologic reports of the incidental mass revealed that grossly, the lesion was a firm, well-circumscribed pseudo encapsulated mass of 4,5 x3x 2,5 cm in dimension; cut section of the tumour showed gray-white to yellow -white colour and fasciculation (Figure-1). Microscopically, the tumour was composed of fibrocollagenous tissue and spindle cells arranged predominantly haphazardly or a short fascicular pattern around thick collagen fibers. The cells had round to oval nuclei and a pale eosinophilic cytoplasm. The degree of cellularity was low and showed keloid like collagen fibers in most areas of the tumour; and mitoses was virtually absent. Immunohistochemically, tumour cells showed a positive reaction for CD34 and vimentin, but a negative reaction for keratin, epithelial membrane antigen (EMA), S-100 protein and desmin (Figure-2). Histology of the nonfunctional kidney was of an atrophic kidney pattern. Based on histologic and immunohistochemical features of the lesion, it was diagnosed as a solitary fibrous tumour.

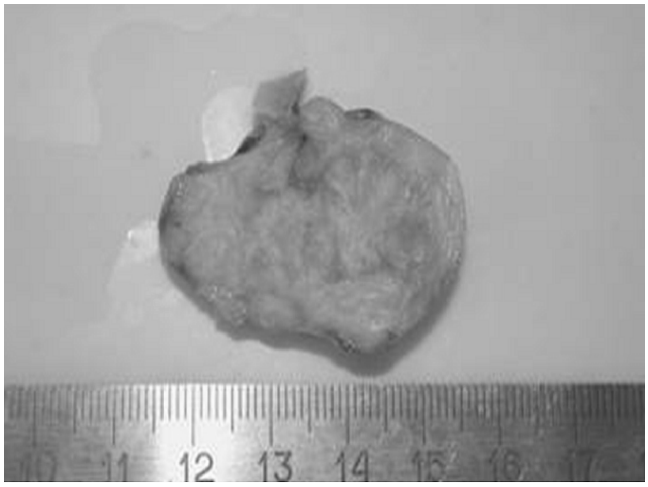


Figure-1: Macroscopic appearance of the cut section of the incidental retroperitoneal solitary fibrous tumour.

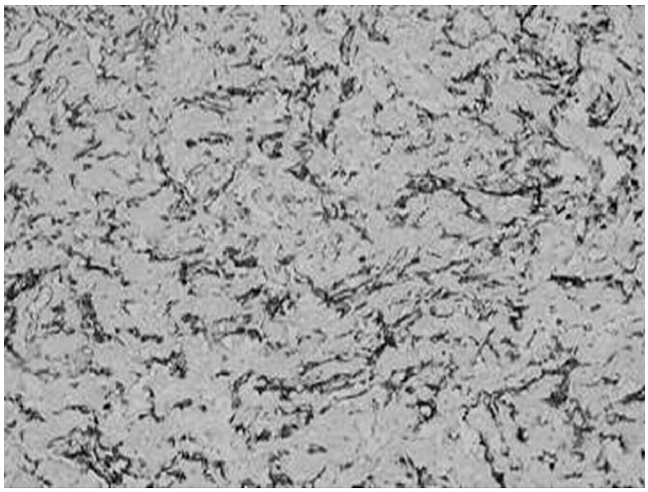

Figure-2: Photomicrograph illustrating CD34 immunopositivity in the neoplastic cells, which is compatible with a diagnosis of solitary fibrous tumour (avidin peroxidase, x100).

The clinical course of our patient was uneventful and subsequent screening showed no further evidence of recurrence or malignancy during one-year period.

## Discussion

Solitary fibrous tumour (SFT) was initially reported by Klemperer and Rabin in 1931 as a mesenchyme derived benign tumour.[7] It usually develops in the pleura and rarely in the retroperitoneum. Nakatani et al have recently reviewed 25 patients (age range 17-82) with solitary fibrous tumours (SFTs) arising in the retroperitoneum.[6] Clinical presentation varies according to the size and localization; including cough, chest pain and dyspnoea, as well as paraneoplastic syndromes such as hypoglycaemia, digital clubbing or swelling in flank with dull pain. In our case, the mild flank pain was a unique symptom that completely recovered postoperatively. The dimensions of reported SFTs vary from 2 to 26 cm in diameter.[6,7] Thus, the lesion in our patient, which was approximately 3 cm in diameter, was a relatively small one. This is probably the reason for not performing an ultrasound. Due to the presence of an atrophic kidney accompanying hypertension, a laparotomy was indicated for a simple nephrectomy. Computerized tomography was not performed preoperatively because of the ultrasound report. SFTs are composed of uniform collagen forming spindle cells which are arranged in interlacing fascicles and show no or minimal

mitotic activity. The vascularity varies from narrow vascular clefts to gaping and branching vascular channels. Prominent vascularity resulting in haemangiopericytoma-like foci is also frequently seen.[8] Most tumours have a variable appearance, with alternating relatively hyper-cellar or hypo-cellar regions. These features suggest a diagnosis of a benign nonepithelial tumour, neurogenic tumour, fibroblastic tumour or haemangiopericytoma. At present, a strong immunoreactivity for CD34 monoclonal antibodies allows the distinction of SFT from most of the other neoplasms.[9] A strong and diffuse immunoreactivity to CD34 and desmin confirmed the diagnosis of SFT in our patient.

It has been reported in SFTs that about 2% of patients with histologically benign tumours, have the possibility of recurrence. Long-term clinical follow-up is recommended for all patients with solitary fibrous tumour. The potential adverse biological behaviour of this tumour, may lead to repeated recurrences or malignant transformation.[10]

The clinical outcome of our patient was favourable and showed no further evidence of recurrence or malignancy during one-year period.

## Conclusion

In conclusion, most extrathoracic solitary fibrous tumours appear to pursue a benign course. Although, some have the potential to recur or metastasize, complete excision and careful long-term follow-up are essential for all patients. Although, it is extremely rare, clinicians should take into account the diagnosis of SFT among the incidental masses of the retroperitoneum especially with ipsilateral atrophic or hypotrophic kidney.

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