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

Murat Savas,¹ Halil Ciftci,² Abdullah Ozgonul,³ Ozgur Sogut,⁴ Muhammet E Guldur²⁵
Department of Urology,¹,² Department of General Surgery,³ Department of Emergency Medicine,⁴ Department of Pathology,⁵ Harran University, School of Medicine, Sanliurfa, Turkey.

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 histopathological 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. 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.

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. 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. 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. 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. Most tumours have a variable appearance,
with alternating relatively hyper-cellular or hypo-cellular 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. 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.

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.

References

1. Witkin GB, Rosai J. Solitary fibrous tumour of the upper respiratory tract. A
2. Young RH, Clement PB, Mc Caughey WT. Solitary fibrous tumours (fibrous
mesotheliomas) of peritoneum: A report of three cases and review of literature.
3. el-Naggar AK, Ro JY, Ayala AG, Ward R, Ordonez NG. Localized fibrous
tumour of the serosal cavities: Immunohistochemical electronmicroscopic and
5. Hasegawa T, Mastsyno Y, Shimoda T, Hirohashi S, Hirose T, Sano T. Frequent
28: 86-91.
6. Nakatani T, Tamada S, Iwai Y, Tanimato Y. Solitary fibrous tumour in the
retroperitoneum: a case with infiltrative growth. Hinyokika Kyio 2002;
48: 637-41.
7. Klemperer P, Coleman PR. Primary neoplasm of the pleura: A report of five
1995; pp 787-819.
9. Westra WH, Gerald WL, Rosai J. Solitary fibrous tumour, consistent CD34
fibrous tumours of the pleura. A clinicopathologic review of 223 cases. Am J