Renal solitary fibrous tumour: A rare pathological entity

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Cite as: *Can Urol Assoc J* 2014;8(9-10):e657-9. http://dx.doi.org/10.5489/cuaj.1854
Published online September 9, 2014.

Abstract

A solitary fibrous tumour (SFT) is a rare mesenchymal cell neoplasm that can develop at any site. SFT of the kidney is extremely rare. Recently, we had a case of solitary fibrous tumour involving the left kidney in a 71-year-old female patient. The SFT was incidentally found by imaging modalities at the time of a physical workup. Computed tomography and retrograde pyelography showed a 4 × 3.5 × 4-cm nodular mass in the middle poles of the left kidney adjacent to the renal pelvis. A laparoscopic radical resection of the left kidney was performed. The tumour was well-circumscribed and composed of a mixture of spindle cells; microscopically, we found dense collagenous bands. Immunohistochemical studies showed strong reactions with CD34, bcl-2 and CD99. A nuclear positivity with Ki-67 was observed in less than 1% of cells. The tumour was negative for desmin, SMA and CD117. Histopathological and immunohistochemical studies confirmed the diagnosis of a solitary fibrous tumour.

Introduction

A solitary fibrous tumour (SFT) is an unusual mesenchymal tumour that is initially recognized in the pleura.[1] An extrapleural SFT, although rare, has recently been discovered in various sites, including the abdomen, retroperitoneum, groin, trunk, thigh, eyelid, orbit, uterine cervix and meninges. Tumours originating from urogenital system organs, such as the kidney, prostate, and urinary bladder, have also been revealed.[2] We report an additional case arising from the left kidney that was successfully managed by laparoscopic surgery. Histopathological and immunohistochemical studies confirmed the diagnosis of a solitary fibrous tumour. We discuss the clinicopathological features, differential diagnosis and treatment of SFT.

Case report

A 71-year-old woman was referred because of a tumour in the left renal found incidentally by imaging modalities at the time of a physical workup at our hospital. She denied a history of hematuria, fever, weight loss or other constitutional symptoms. Physical examination and laboratory data were unremarkable. Abdominal ultrasonography demonstrated a solid, relatively well-demarcated, hypoechoic mass, about 3.2 × 3.8 cm, occupying the left renal pelvis. A computed tomography (CT) showed a 3.1 × 4.1-cm, heterogeneous and poorly enchancing mass in the left renal pelvis, slightly compressing the collecting system outwards (Fig. 1). Retrograde pyelography showed a slight dilation and deformation of the left renal pelvis calyces. Small filling defect were seen in the left renal pelvis. The left ureter did not have any abnormalities. Cystoscopy showed no abnormalities in the urinary bladder. Chest x-ray, chest CT scan and a bone scans were all negative for metastasis. Based on the radiological findings, laparoscopic radical resection of the left kidney was performed to remove the tumour.

Macroscopic examination of the 10 × 5.5 × 5.5-cm nephrectomy specimen revealed a 4 × 3.5 × 4-cm nodular mass in the middle poles of the left kidney adjacent to the renal pelvis. Resection margins were free of tumour and there was no invasion into perinephric adipose tissue and all evaluated lymph nodes were negative. The cut section of the mass was gray-white in colour and hard in consistency. Histologic examination of the tumour showed spindle-shaped cells arranged in short fascicles with abundant thick bundles of hyalinized collagen. Cytological atypia or mitotic figures were not identified. The normal renal parenchyma was focally affected (Fig. 2). Immunohistochemically, the tumour cells were diffusely positive for CD34 (Fig. 3, part A), CD99 (Fig. 3, part B), bcl-2 (Fig. 3, part C) and focally for Caldesmon (Fig. 3, part D). However, staining for desmin, SMA and CD117 were negative. A nuclear positivity with Ki-67 was observed in less than 1% of cells.

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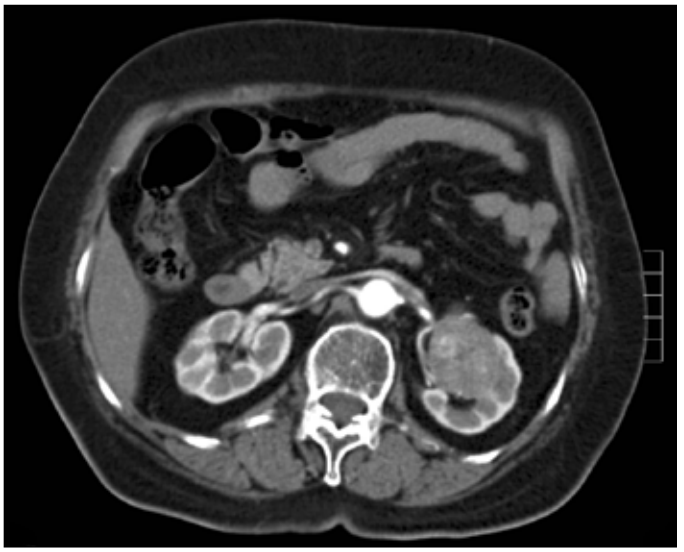

Fig. 1. The computed tomography scan shows a well-circumscribed homogeneous tumour invading the left renal pelvis; the mass is poorly enhanced on contrast.

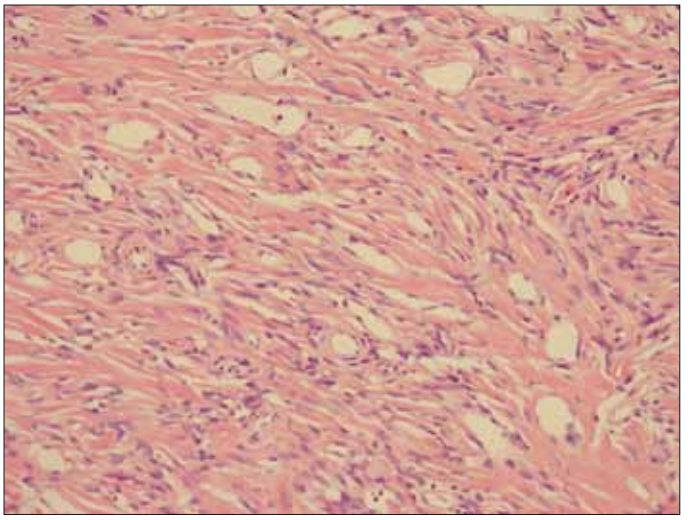

Fig. 2. Fascicular arrangement of spindle cells with uniform, oval, vesicular nuclei without atypia (Hematoxylin and eosin stain, ×200).

Based on the histopathological and immunohistochemical findings, a diagnosis of a solitary fibrous tumour was established. Because of the patient's advanced age and the final diagnosis, chemotherapy and radiation therapy were not given postoperatively. The patient was discharged 10 days after surgery. No recurrence was observed after 2 years.

Discussion

SFT is a rare type of spindle cell neoplasm originating from mesenchymal cells and usually arises in the pleura.[3] However, it is not anatomically restricted to the chest cavity and has been described in various extrapleural sites, such as upper respiratory system, lung, liver, nasal cavity, paranasal sinuses, mammary glands, orbita, mediastinum, greater salivary glands, meninges, and even the kidney.[4-8]

The most common clinical symptom of SFT of the kidney is the typical triad of flank pain, accompanied with gross hematuria or a palpable abdominal mass. In our case, the patient's condition was found incidentally by imaging modalities at the time of a physical workup with no symptoms. However, based on the clinical and imaging studies, it was difficult to make the differential diagnosis from other primary benign and malignant monomorphous spindle cell tumours of the kidney. But the final diagnosis of a SFT can be made by means of pathology. Microscopically, SFT is characterized by long spindle cell proliferation showing a patternless architecture with a combination of alternating hypocellular and hypercellular areas separated from each other by thick bands of hyalinized, somewhat keloidal collagen and branching hemangiopericytoma-like vessels.[9] Cytoplasms of the spindle cells appear scarce and ill-bordered. In most benign cases, mitotic activity and atypia have not been observed.[8]

With regard to the immunohistochemical study, SFT has been shown to be highly positive for CD34, CD99, and bcl-2, and CD34 has been considered as a specific immune- peroxidase marker for SFT.[10-13] Other immunohistochemistries involving cytokeratin, SMA, S-100 protein, desmin, CD117 and epithelial membrane antigen are negative, but useful for differential diagnosis of SFT. Therefore, we diagnosed this case as a SFT based on the following histological features: the tumour cells were diffusely and strongly positive for CD34, and a positive expression of bcl-2, CD99 and Caldesmon was also present in this case. Staining for desmin, SMA and CD117 were negative.

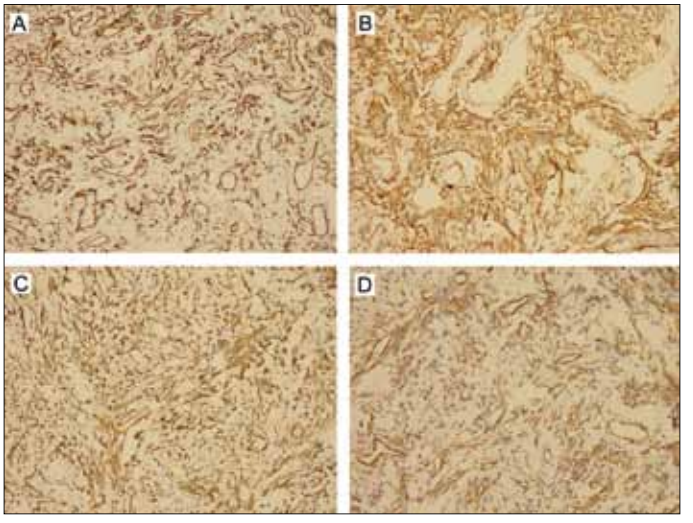

Fig. 3. Immunohistochemistry of the solitary fibrous tumour of the kidney. The tumour cells were diffusely positive for CD34 (A), CD99 (B), bcl-2 (C) and focally for caldesmon (D) (immunohistochemistry, original magnification, ×200).

The treatment of a SFT is surgical removal. Although most SFTs of the kidney exhibit benign behaviour, excision is recommended to establish a diagnosis, alleviate any symptoms, and prevent documented risk of malignant transformation.[14] Laparoscopic excision has been widely used, and is considered the first-line surgical technique.[12,15] In this case, preoperative examination showed a 4 × 3.5 × 4-cm mass in the middle poles of the left kidney adjacent to the renal pelvis, and a laparoscopic radical nephrectomy was performed.

Conclusion

A SFT of the renal pelvis is a rare, mesenchymal cell tumour with a favourable prognosis. The combination of clinical, biochemical and radiological features may help in lesion characterization, but only histology can provide the definite diagnosis. Radical resection is the first choice of treatment. Given that malignancy, recurrence or both have been reported in up to 10% to 15% of cases, a longer follow-up period might be necessary to definitively evaluate the clinical outcome of a renal SFT.

Competing interests: Biao Dong, Jianjian Zhang, Gang Wang, Xiuyu Zhai, Yaowen Fu, Honglan Zhou and Yuantao Wang all declare no competing financial or personal interests.

This paper has been peer-reviewed.

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