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CASE REPORT

Renal Solitary Fibrous Tumor - A Case Report

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ABSTRACT

A renal solitary fibrous tumor (SFT) is an unusual and especially rare entity. Preoperative diagnosis is usually difficult, but it remains of favorable prognosis. In this study, the authors report a case of a 33-year-old Moroccan man with a history of left pyelolithotomy who presented with a 19 cm left renal mass associated with two kidney stones in the lower calyx. He underwent an open radical nephrectomy, and pathology revealed a renal SFT. To the best of our knowledge, our patient is the first case of renal SFT with a history of urolithiasis and presenting with concomitant ipsilateral calculi. Radical nephrectomy is still the cornerstone gold standard therapy for patients with large renal tumors, especially for rare tumors with no proper protocol.

KEYWORDS: Solitary fibrous tumor, Renal, Open radical nephrectomy

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INTRODUCTION

A renal solitary fibrous tumor (SFT) is an unusual and especially rare entity [1]. Preoperative diagnosis is usually difficult [2], but the prognosis remains favorable. There are very few reports on SFT originating in the kidney, and, to the best of our knowledge, our patient is the first case of renal SFT with a history of urolithiasis and presenting with concomitant ipsilateral calculi.

CASE REPORT

A 33-year-old Moroccan man with a history of left pyelolithotomy was referred to our hospital for persistent left flank pain. He had not had any hematuria, urgency, frequency, or dysuria. However, a huge abdominal mass was discovered on physical examination. Abdominal computed tomography (CT) showed a well-limited left renal mass of 20 cm in diameter that enhanced heterogeneously due to necrosis areas. The tumor involved the renal artery and vein, and the CT also revealed two kidney stones of 8 and 6 mm in the lower calyx (Figure 1 A, B). Blood and urine laboratory data, including cytology, were within normal limits. The diagnosis was a malignant renal tumor of the left kidney based on the CT results. The patient underwent an open radical nephrectomy, and the tumor had a pseudocapsule that made it feasible to be dissected with no residual tumor.

Gross examination showed a 911-gram, 19 x 15 cm, capsule-coated, solid mass. Microscopically, spindle cells were seen with some ischemic necrosis areas. The cytoplasm was moderately abundant, and there was rare nuclear atypia. Six mitoses per 10 high-power fields were found in some areas, and the tumor focally infiltrated perirenal fat.

Immunohistochemical staining was positive for cluster of differentiation (CD) 34, Ki67 (about 10% positive), vimentin, and smooth muscle actin. Nevertheless, stainings for S100 polyclonal antibody, CD31, human melanoma black 45 (HMB-45), and cytokeratin were negative (Figure 2 A, B).

The patient did not receive any adjuvant chemotherapy before and after surgery. No complication was detected during the perioperative period.

The patient had no evidence of recurrences or metastases on CT six months after surgery (Figure 3).

DISCUSSION

SFT is a mesenchymal spindle cell tumor that most commonly occurs in the pleura, but it can also be found in extrapleural locations [3].

Renal SFT is especially rare, with less than 100 cases reported in the English literature [1,2,4]. The first case was described by Gelb et al. in 1996 [5]. According to the WHO classification of kidney tumors, it is a "fibroblastic mesenchymal tumor with malignant potential" [2]. The renal capsule is the most common site of origin of renal SFT [3,6]. There is no gender predominance, and the mean age is 52 years [2].

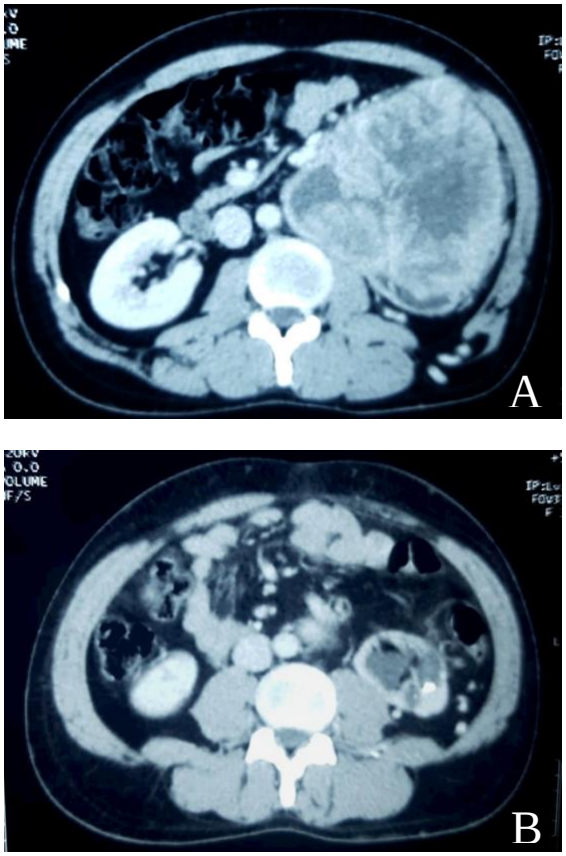

Figure 1: Preoperative CT Scan.

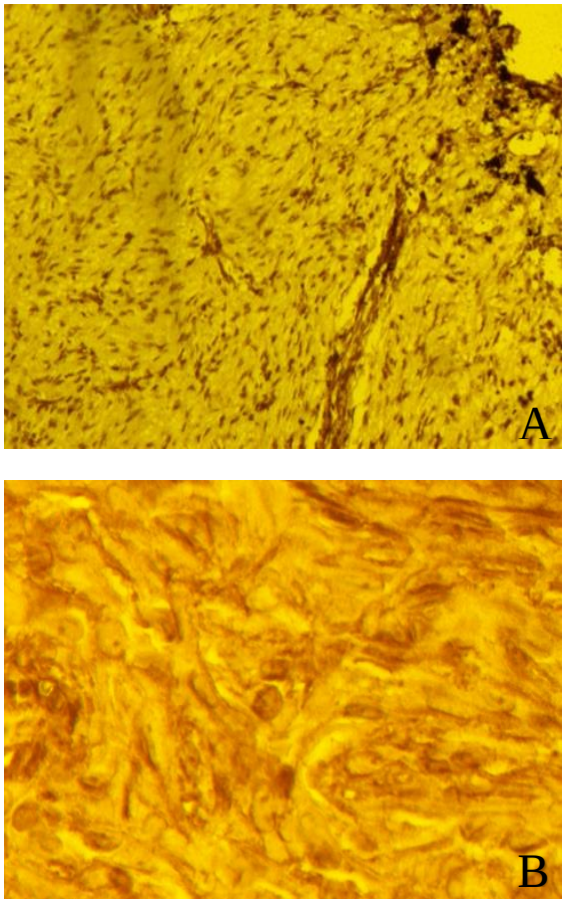

Figure 2 : Immunohistochemical staining
A: Smooth muscle anti-actin antibody (Clone 1A4, Dako).
B: Anti-CD34 antibody (Clone QBEnd-10, Cell Marque).

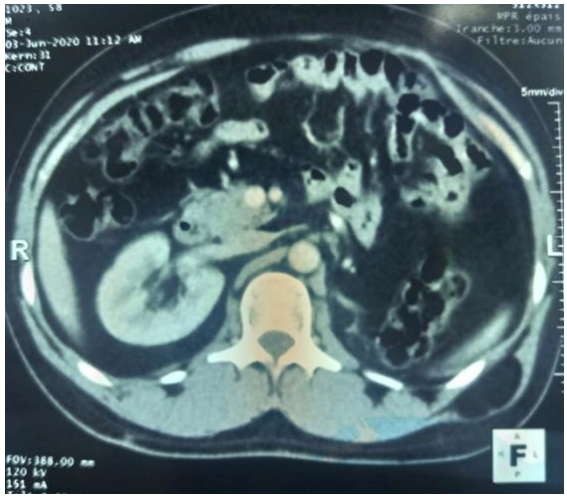

Figure 3: Postoperative CT Scan.

Renal SFT presentation is nonspecific and includes abdominal pain, gross hematuria, and a palpable mass, mimicking renal cell carcinoma (RCC) [2]. However, unlike RCC, renal SFTs rarely cause paraneoplastic syndromes [3] and are often incidentally diagnosed on imaging examinations [1].

Like our patient, patients with a history of kidney stones are exposed to an increased risk of RCC and upper tract urothelial carcinoma (UTUC) [7]. However, there appears to be no link between SFT and urolithiasis.

Renal SFTs present as a hypo- or heterogenous echogenic mass with relatively well-defined margins [2,3]. CT scans are more accurate in diagnosing these tumors, showing a strong enhancement associated with cysts and areas of hemorrhage or necrosis that can confirm the diagnosis [3]. MRI is indicated in case of enhanced CT scan contraindication or uncertainty [3].

Renal SFTs are almost benign tumors [3]. Microscopic findings show irregular beam- or spindle-shaped vortex cells [2]. The immunohistochemical examination is crucial in determining the diagnosis; the tumor cells of SFT are diffusely positive for CD34 (considered as a specific immune-peroxidase marker for SFT [3]), CD99, and Bcl-2 [2]. Rarely, the histopathological examination could show increased cellularity, pleomorphism, presence of hemorrhage, necrosis, sarcomatous overgrowth, increased mitotic activity (>4 mitoses per 10 high-power fields), and a 20% Ki67 proliferative index, suggesting malignancy [3,8]. However, the large size of the tumor may result in the absence of malignant features in the entire tumor tissue [4].

The progression of untreated renal SFT is not documented due to its rarity, and preoperative diagnosis is usually difficult [2]. However, it has a low risk of metastasis [2], with only 10% of extrathoracic SFTs recurring or metastasizing [3].

Radical nephrectomy is the primary treatment for both benign and malignant renal SFTs [2]. There is no evidence that partial nephrectomy has the same oncological outcome as radical nephrectomy [1,3].

Clinical behavior is unpredictable [2], but some prognostic factors of metastasis and death are identified: tumor size (>15 cm), age (>55 years), and mitotic counts (>4 high-power fields) [9].

Furthermore, long-term follow-up is necessary for all patients [2].

CONCLUSION

In patients with large renal tumors, radical nephrectomy is still the cornerstone gold standard management of kidney tumors, especially in rare tumors where there is no proper protocol. In renal SFTs, the diagnostic criteria for malignant SFTs are purely microscopic, and long-term follow-up is required even in the absence of malignant findings.

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None.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript, and provided approval for this final revised version.

COMPETING INTERESTS

The author declares no competing interests with this case.

PATIENT CONSENT

Written informed consent was obtained from the patient for the publication of this case report

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