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(CASE REPORT)

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# Primary fibrosarcoma of kidney in an adult: A diagnostic and therapeutic challenge: A case report and literature review

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## Abstract

**Background:** Primary fibrosarcoma of kidney in adults is a rare entity with a few cases described in the literature. This article will focus on an atypical presentation of this tumor.

**Summary**: We report a case of a 57-year-old male with a 3-year history of intermittent left flank pain, without hematuria or other significant symptoms. Imaging studies, including CT, revealed a large, well-circumscribed mass in the left kidney, initially diagnosed as renal cell carcinoma. The patient underwent a left radical nephrectomy, and histopathological examination revealed a spindle cell proliferation with areas of central necrosis. Immunohistochemistry initially suggested a diagnosis of renal angiomyolipoma. However, six months later, the patient presented with recurrent left-sided lumbar pain and metastases. A re-evaluation of the initial nephrectomy specimen and a biopsy of the retroperitoneal mass confirmed the diagnosis of primary renal fibrosarcoma. The patient is currently receiving chemotherapy.

**Conclusion**: This case illustrates the diagnostic challenges and aggressive nature of primary renal fibrosarcoma. Despite radical nephrectomy, the patient developed rapid recurrence and metastasis, underscoring the poor prognosis of this rare tumor. Further research is needed to establish effective treatment protocols, as current approaches are based primarily on case reports.

**Keywords:** kidney; Renal fibrosarcoma; Radical nephrectomy; Chemotherapy

## 1. Introduction

Renal sarcomas are extremely rare, accounting for approximately 1 to 3% of malignant renal tumors [1]. Histopathologically, renal sarcomas are malignant mesenchymal tumors of the kidney, and among them, primary renal fibrosarcoma is one of the rarest types. Other renal sarcomas include leiomyosarcoma, rhabdomyosarcoma, and liposarcoma.

Renal fibrosarcoma generally develops from the renal capsule, which is rich in fibrous and connective tissue, explaining its origin within mesenchymal renal tumors. Although renal fibrosarcomas can occur at any age, they are more common in young and middle-aged adults. Their clinical presentation varies, ranging from abdominal pain to more specific symptoms such as hematuria, which is often the hallmark of the disease.

The diagnosis of fibrosarcoma is a diagnosis of exclusion, as the absence of specific immunological markers for fibroblasts complicates management [2]. To date, no standard treatment exists for patients with renal fibrosarcoma.

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Radical nephrectomy remains the primary therapeutic approach. Differential diagnosis is complex, as renal fibrosarcoma must be distinguished from more common renal neoplasms, such as renal cell carcinoma.

## 2. Case Report

A 57-year-old male, with no significant medical history or toxic habits, presents with intermittent left flank pain evolving over the past 3 years, without macroscopic hematuria or other associated symptoms. He is in overall good health, without fever. Clinical examination reveals a firm, painless palpable mass in the left flank, measuring approximately 16 x 12 cm. Routine blood tests, including a complete blood count, renal function tests, and cytobacteriological examination of urine, are within normal limits.

Contrast-enhanced abdominopelvic CT scan show an enlarged left kidney, measuring 20.5 cm x 6.1 cm, with a lower polar tumor mass. The tumor is roughly oval, encapsulated, with central necrotic areas and no microcalcifications. The tissue portion is homogeneously enhanced after contrast, measuring approximately 13 cm x 11.3 cm x 12.9 cm. A slight infiltration of the left perirenal fat is noted, without extension beyond Gerota's fascia, and no extension into the renal vein or inferior vena cava (Figure 1). A clinical diagnosis of renal cell carcinoma is made, and the patient undergoes a left radical nephrectomy. The postoperative period is uneventful.

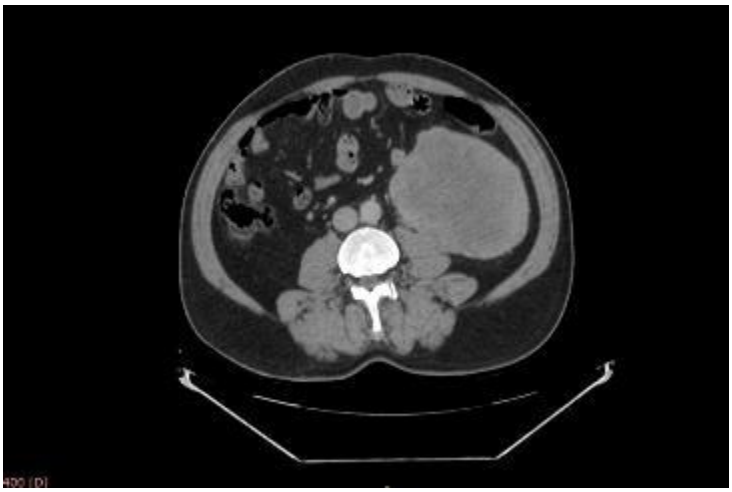

**Figure 1** Axial contrast-enhanced CT image showing a large, encapsulated lower pole left renal mass with central necrosis and mild perirenal fat infiltration, without vascular or extracapsular extension

Macroscopic examination of the nephrectomy specimen revealed a well-circumscribed tumor weighing 2 kg and measuring 25 x 17 x 11 cm. The tumor was located 1 mm from the renal capsule, exhibiting a solid, homogeneous beige appearance, with no macroscopic evidence of necrosis or hemorrhage. Invasion of the excretory pathways, calyces, renal pelvis, and renal hilum was noted, without signs of vascular thrombosis. The tumor was found to be 0.5 cm from the vascular resection margin at the renal hilum.

Microscopic examination revealed a low-cellularity spindle cell proliferation arranged in diffuse sheets. The nuclei were monomorphic, ovoid to spindle-shaped, with finely nucleolated granular chromatin. The cytoplasm was eosinophilic, abundant, and showed indistinct borders. The stroma was hemangiopericytoma-like, composed of dilated, thin-walled vascular structures, associated with extensive areas of central coagulative necrosis estimated at 60%, with no viable cells identified.

Immunohistochemical analysis was performed, revealing negative staining for anti-Melan A, anti-HMB-45, and anti-Bcl2 antibodies, supporting the diagnosis of renal angiomyolipoma.

Six months later, the patient presented with a recurrence of left-sided lumbar pain, associated with asthenia, anorexia, and a 10-kg weight loss. A thoraco-abdominopelvic computed tomography (CT) scan revealed a large mass in the left renal fossa, measuring 22 × 10 cm, with areas of necrosis and invasion of the psoas muscle. A 34-mm pre-aortic lymphadenopathy was noted, causing right-sided uretero-pyelocaliceal dilation (Figure 2). Additional findings included pulmonary nodules, a hepatic lesion, and peritoneal carcinomatosis, all suggestive of metastatic spread.

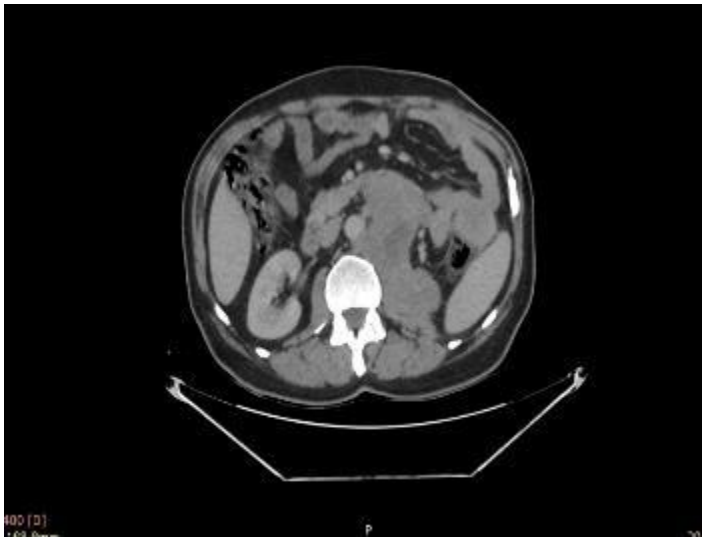

**Figure 2** Axial contrast-enhanced CT image showing a large heterogeneous mass in the left renal fossa with areas of necrosis and invasion of the left psoas muscle

The patient underwent right ureteral stent placement and a biopsy of the retroperitoneal mass, along with immunohistochemical re-evaluation of the initial left nephrectomy specimen. Morphological and immunohistochemical findings confirmed the diagnosis of a left renal fibrosarcoma. The patient is currently receiving systemic chemotherapy based on doxorubicin and ifosfamide.

## 3. Discussion

Renal sarcomas are rare tumors, accounting for approximately 1% to 3% of malignant renal neoplasms [3]. Among them, primary renal fibrosarcoma is exceptionally rare, with only six cases reported in the literature to date [5, 6, 7].

Renal fibrosarcomas predominantly occur in patients aged between 40 and 60 years, affecting both males and females. The right kidney is more commonly involved; however, in our case, the tumor was located on the left side. The tumor size is usually significant by the time medical consultation is sought. It is important to note that these observations are based solely on the currently available case reports [7].

Recent advances in immunohistochemical techniques have facilitated a more accurate distinction of true cases of primary renal fibrosarcoma. Cavaliere et al. [8] reported one case in a 10-year review of primary renal sarcomas, while Grignon et al. [4] identified one case in a series of 17 primary renal sarcomas.

To establish the diagnosis of a primary renal sarcoma, three essential criteria must be met: 1) the patient must have no history or evidence of sarcoma elsewhere, to rule out metastatic disease; 2) the macroscopic appearance must support a renal origin, rather than retroperitoneal sarcoma infiltration; and 3) sarcomatoid renal cell carcinoma must be excluded [4].

Most cases of renal fibrosarcoma are challenging to diagnose at an early stage due to the absence of specific symptoms. Common clinical features, such as an abdominal mass, hematuria, and flank pain, typically appear at more advanced stages. In some instances, the diagnosis is incidental, as in our case, where the patient presented with intermittent left-sided lumbar pain, without macroscopic hematuria or associated symptoms, in an afebrile context with preserved general condition.

Benign mesenchymal renal masses, such as angiomyolipomas, are relatively common; however, their malignant counterparts—such as leiomyosarcomas and fibrosarcomas—are exceedingly rare. No imaging modality can reliably differentiate these various subtypes.

Computed tomography (CT) typically reveals solid renal masses of variable size, often measuring several centimeters in diameter, with either well-defined or infiltrative borders. Contrast-enhanced CT may demonstrate heterogeneous

enhancement and central necrotic areas. Venous extension has been described in approximately half of the reported cases in the literature [9].

In our case, the contrast-enhanced CT scan revealed a large lower pole renal mass with subtle infiltration of the perirenal fat, without evidence of venous extension. Magnetic resonance imaging (MRI) may also aid in the diagnosis of renal fibrosarcoma, with lesions typically demonstrating hyperintensity on T1-weighted images and heterogeneous signal intensity on T2-weighted sequences [10].

Histologically, typical renal fibrosarcoma presents with fusiform cells arranged in parallel rows, intersecting at acute angles, and areas of hemorrhage and necrosis. The cells have weak to moderate eosinophilic cytoplasm, and the nuclei are oval with irregularly distributed chromatin [11].

Immunohistochemistry (IHC) is a biochemical technique used to identify specific molecules in different types of tissues. The tissue is treated with antibodies that bind to the specific molecule, which are then made visible under a microscope through a color reaction, radioisotope, or fluorescent dye. Mesenchymal masses such as fibrosarcoma, leiomyosarcoma, and sarcomatoid renal carcinoma can appear similar in histopathology. Immunohistochemistry is the definitive method to exclude the latter two, as fibrosarcoma is typically a diagnosis of exclusion. Fibrosarcomas are positive for vimentin (non-specific), and negative for cytokeratin, desmin, and HMB-45 (Human Melanoma Black 45), whereas sarcomatoid renal carcinoma and leiomyosarcoma are diffusely positive for cytokeratin and desmin, respectively [11]. In our case, the immunohistochemical study revealed the absence of staining for anti-desmin, anti-cytokeratin, and anti-HMB-45 antibodies, and the anti-vimentin antibody was not tested.

To date, only six cases of primary renal fibrosarcoma in adults have been reported in the literature. In all of these cases, management consisted of radical nephrectomy, often justified by the local aggressiveness and extent of the tumor at diagnosis. Adjuvant chemotherapy was administered in each case.

Radical nephrectomy remains the gold standard for treating primary renal fibrosarcoma. Therapeutic management mainly relies on individualized approaches, and no established consensus exists due to the small number of reported cases.

Despite the absence of metastases at diagnosis, renal fibrosarcoma has a poor prognosis, with a 5-year overall survival rate of less than 10% [12]. Chemotherapy and radiotherapy have shown no significant effect. A recent study showed that soluble long pentraxin 3 (PTX3) receptor may act as an oncosuppressor by functioning as an antagonist to the fibroblast growth factor (FGF)/FGF receptor (FGFR) pathway to inhibit FGF-dependent tumor growth [13].

Jain et al. [14] found that miR-197-3p can significantly inhibit viability, colony formation, and migration of fibrosarcoma cells, while inducing G2/M cell cycle arrest.

For recurrent fibrosarcoma with high vascular endothelial growth factor (VEGF) expression, apatinib may effectively reduce the risk of disease progression in patients with recurrent fibrosarcoma with high VEGF expression [15].

These findings offer new perspectives for the treatment of renal fibrosarcoma.

## 4. Conclusion

Primary renal fibrosarcoma remains one of the rarest forms of renal malignancy, posing significant diagnostic and therapeutic challenges. Due to its nonspecific clinical presentation and lack of distinctive radiologic or immunohistochemical features, diagnosis often relies on careful exclusion of other renal tumors. Surgical resection through radical nephrectomy remains the mainstay of treatment; however, the high rate of local recurrence and distant metastases underscores the aggressive nature of this tumor and its poor prognosis. Given the limited number of reported cases, there is currently no consensus on optimal adjuvant therapy. Advances in molecular profiling and targeted therapies may pave the way for more effective treatment options in the future, highlighting the need for further research and collaborative case reporting.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

The authors report no competing personal or financial interest related to this work..

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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