A Rare Case of Primary Fibrosarcoma of Kidney

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Primary sarcomas of kidney are rare tumors accounting for 1% to 3% of all primary renal malignancies. Among sarcomas fibrosarcoma is rare. Here we report a case of primary fibrosarcoma of the kidney in a 70-year-old man who presented with gradually increasing abdominal swelling and pain.

Keywords. kidney, fibrosarcoma, vimentin

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INTRODUCTION

Primary renal malignancies are mainly renal cell carcinoma and transitional cell carcinoma. Primary renal fibrosarcoma is a very rare tumor. Whenever it is diagnosed, it needs to be differentiated from other more common spindle cell lesions, such as leiomyosarcoma and sarcomatoid renal cell carcinoma. With the help of immunohistochemical markers, nowadays primary renal fibrosarcoma is easily differentiated from its alike tumors. Cytokeratin, desmin, and vimentin are the immunohistochemical markers which help to categorize the lesion. Here we report a case of primary fibrosarcoma of the kidney in a 70-year-old man.

CASE REPORT

A 70-year-old man, nonsmoker and alcoholic, presented with complaints of gradually increasing abdominal lump and pain in the right flank region with the constitutional symptoms of fever, anorexia, and weight loss. General examination revealed a palpable firm lump in the right flank region, about 17 × 12 cm in size. The liver and spleen were not palpable. Routine blood analysis including complete blood count, kidney function test, and urinalysis were within normal limits. Ultrasonography and contrast enhanced computed tomography of the abdomen showed a heterogeneously enhanced lobulated mass in the right kidney measuring 17 × 10 cm. No extension in the renal vein or in inferior cava was noted. A clinical diagnosis of renal cell carcinoma was made.

The patient underwent right radical nephrectomy, and the postoperative period was uneventful. Grossly, the specimen of the right kidney was measuring 17 × 10 × 6 cm with bosselated external surface (Figure 1). Cut surface showed a lobular grayish white tumor involving almost the whole kidney and reaching up to the capsule. The ureter was not involved. Multiple sections were taken for histopathological examination. Microscopic examination revealed partially encapsulated tumor tissue comprising of spindle cells in fasicular arrangement. The cells were spindle to round in shape with scant to moderate eosinophilic cytoplasm. The nuclei were spindle to oval with irregular nuclear membrane and irregularly distributed chromatin. Cells were showing moderate anisonucleosis (Figures 2 and 3). Based on these morphological findings, a differential diagnosis of sarcomatoid renal cell carcinoma and fibrosarcoma

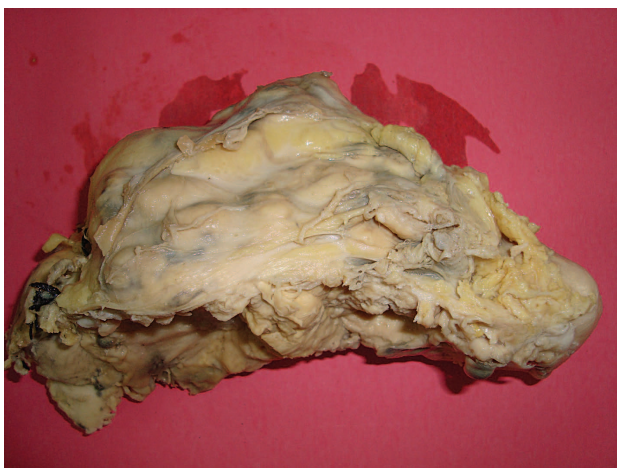

Figure 1. Specimen of the right kidney measuring 17 × 10 × 6 cm with bosselated external surface.

Case Report

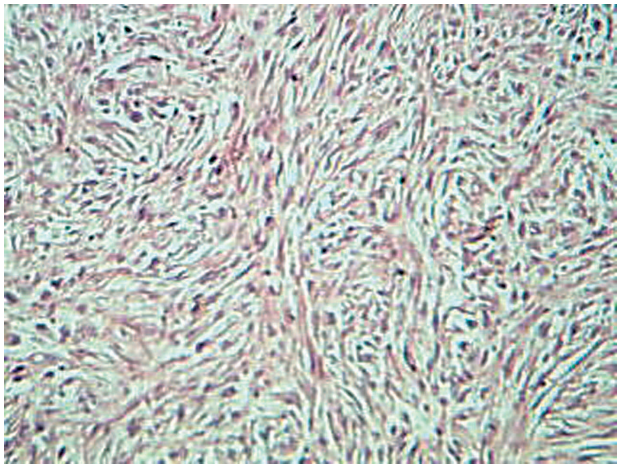

Figure 2. Spindle cells in fasicular arrangement (× 10).

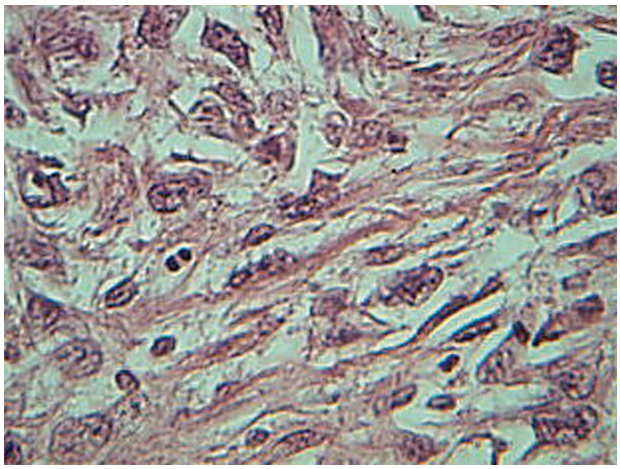

Figure 3. Spindle to round cells with scant to moderate eosinophilic cytoplasm spindle to oval nuclei irregular nuclear membrane showing moderate anisonucleosis (× 40).

of kidney was considered.

Immunohistochemical staining was done with a panel of markers, including pancytokeratin, desmin, and vimentin. The tumor cells were negative for most of the markers except for vimentin, which was diffusely positive in most of the tumor cells (Figure 4). Based on the morphology and immunohistochemical staining, a final diagnosis of fibrosarcoma of the kidney was made.

DISCUSSION

Primary sarcomas of kidney are a rare entity contributing up to 1% to 3 % of all renal tumors.[1,2] The majority of primary sarcomas of kidney are leiomyosarcomas, and rarely, they are fibrosarcomas. Previously, in the nonimmunohistochemical era, all such cases were grouped under one common

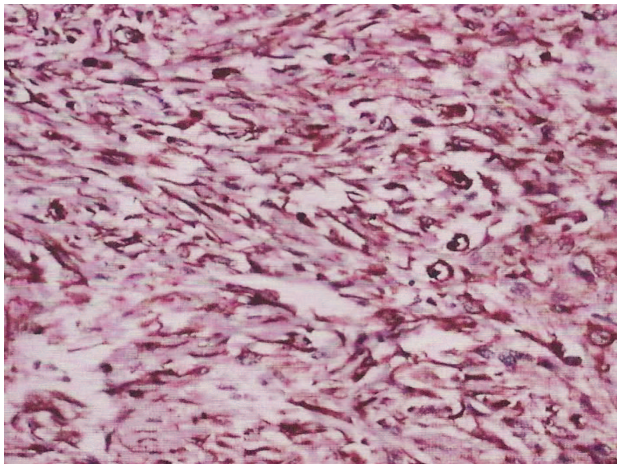

Figure 4. Vimentin positivity on immunohistochemistry.

roof of sarcoma. Some reported cases were actually sarcomatoid renal cell carcinomas, proved with the help of immunohistochemical staining of the retrospective cases.[3,4] Nowadays, with the availability of improved immunohistochemical technique, pathologists are able to separate the actual number of primary fibrosarcoma of the kidney from sarcomatoid renal cell carcinoma. Cavaliere and colleagues found a single case in a 10-year survey of primary renal cell sarcomas, and Grignon and coworkers found a single case in a study of 17 cases of primary sarcomas of the kidney.[3,4] These can be differentiated only on the basis of immunohistochemistry, as fibrosarcomas are diffusely positive for vimentin and negative for cytokeratin and desmin, whereas sarcomatoid renal cell carcinomas and leiomyosarcomas are positive for cytokeratin and desmin, respectively.

Primary renal fibrosarcoma is a very rare malignancy of the kidney. Fibrosarcoma of the kidney usually develops from the capsule of the kidney, which contains fibrous and connective tissue. Fibrosarcoma is commonly seen at the ages of 40 to 60 years with equal sex ratio. Our case, however, was a 70-year-old man. Because of lack of significant symptoms these, tumors are difficult to diagnose early. The usual presenting symptoms are abdominal mass, pain, and hematuria. Some patients are diagnosed when gastrointestinal or metastatic symptoms develop.[5] The tumor is large, solid, and fleshy with infiltrative margins.

Radial nephrectomy seems to be the only treatment for renal fibrosarcoma. Five-year survival rates after surgery are poor and have been reported

to be less than10%.[6] Pettirssen, in his review of 21 cases, found only 2 long-term survivors.[7] The World Health Organization's classification of renal carcinoma categorizes renal tumors into renal cell, metanephric, mesenchymal, mixed, neuroendocrine, and germ cell. Renal cell carcinoma arises from lining epithelium of the kidney tubules and accounts for approximately 90% of renal tumors. Renal cell carcinomas are further classified into clear cell, papillary, and chromophobe, any of which variant can have sarcomatoid changes.

Renal cell carcinoma is generally a tumor of adults with male predominance. All microscopic type of renal cell carcinoma shows reactivity for epithelial markers such as keratin. Co-expression of keratin and vimentin is the rule. The second most common tumor of the kidney is transitional cell carcinoma. Transitional cell carcinoma arises from the renal pelvis. Transitional cell carcinomas are linked with cigarette smoking and occupational exposure to carcinogens.

In conclusion, primary fibrosarcoma of the kidney is a rare, but an aggressive malignant renal tumor, with a very poor prognosis and a low 5-year survival. Hence, it must be differentiated from its morphological look-alikes, leiomyosarcomas and sarcomatoid renal cell carcinomas by immunohistochemistry. In order to make a diagnosis of primary fibrosarcoma of the kidney, one must be sure that patient does not have or had sarcoma elsewhere to exclude the metastatic disease.

CONFLICT OF INTEREST

None declared.

REFERENCES

1. Granmayeh M, Wallace S, Barrett AF, Fisher R, Heslep JH. Sarcoma of the kidney, angiographic features. Am J Roentgenol. 1977;129:107-12.
2. Srinivas V, Sogani PC, Hajdu SI, Whitmore WF Jr. Sarcomas of the kidney. J Urol. 1984;32:13-16.
3. Cavaliere A, Fratini D, Legitimo C, Tosi F, Lolli E. Renal fibrosarcoma: Single cell in a ten year survey. Pathologica. 1984;76:615-2.
4. Grignon DJ, Ayala AG, Ro JY, El-Naygar A, Papadopoulos NJ. Primary sarcoma of the kidney: A clinicopathologic and DNA flow cytometric study of 17 cases. Cancer. 1990;65:1611-8.
5. Kansara V, Powell I. Fibrosarcoma of kidney. Urology. 1980;16:419-21.
6. Judd ES, Donal JM. Sarcoma of the kidney of the adult: A review of twenty cases with a report of a case. Ann Surg. 1931;96:1028-35.
7. Pettirssen RO. Mesenchymal neoplasms of kidney. In: Petersen RO, editor. Urologic pathology. Philadelphia: JB Lippincott; 1986. p. 113.

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