Case Report
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Primary Renal Angiosarcoma

Osmar E. Souza, Renata M. Etchebehere, Marcus A. Lima, Paulo R. Monti

Section of Urology and Pathology, Triangulo Mineiro School of Medicine, Uberaba, Minas Gerais, Brazil

ABSTRACT

Primary renal Angiosarcoma is a rare neoplasm and only 24 cases have been reported in specialized literature.
We describe a case of primary renal angiosarcoma in a patient presenting with hematuria, palpable abdominal mass, left flank pain and anemia. A computerized tomography of the abdomen with contrast medium showed a tumor with 15 cm diameter, in the upper pole of the left kidney, with a low-density central area, suggesting necrosis or hemorrhage. Diagnosis was given in a morphologic base and proven by an immunohistochemical study.
Primary renal angiosarcoma should be included among differential diagnosis of retroperitoneal hematoma and hemorrhagic renal tumors.

***Key words**: kidney neoplasms; angiosarcoma; nephrectomy; hematoma; retroperitoneal space*
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INTRODUCTION

Sarcomas constitute 2 to 3% of primary malignant renal tumors and leiomyosarcoma corresponds to 60% of them (1).

Angiosarcomas are localized or multicentric tumors, with various grades of differentiation and are originated in the endothelium of the blood and lymphatic vessels (1,2). They are high degree malignant neoplasms of primary occurrence, extremely rare in the kidneys, with 24 cases described (3). They are frequently hemorrhagic tumors, being able to simulate a retroperitoneal hematoma or cause massive hematuria (1-3).

We present a case of primary renal angiosarcoma and emphasize its hemorrhagic behavior.

CASE REPORT

A 75 year-old black man, with 3 episodes of macroscopic hematuria, left flank pain, emaciation of 5 k, somnolence and hyporexia starting 1 month before. He presented himself colorless and with a hard mass, little mobility, that extended until 10 cm bellow the left costal edge. Hemorrhage was 6.7 g/dL (13.3 a 17.7) and hematocrit 19.3 % (40 to 52). Abdomen computerized tomography with endovenous contrast medium showed an heterogeneous mass, with an hypodense central area and a peripheral enhancement measuring 15 x 13 x 10 cm, in the superior pole of the left kidney suggesting a renal tumor (Figura-1). Other abdominal organs and the radiography of the thorax were normal.

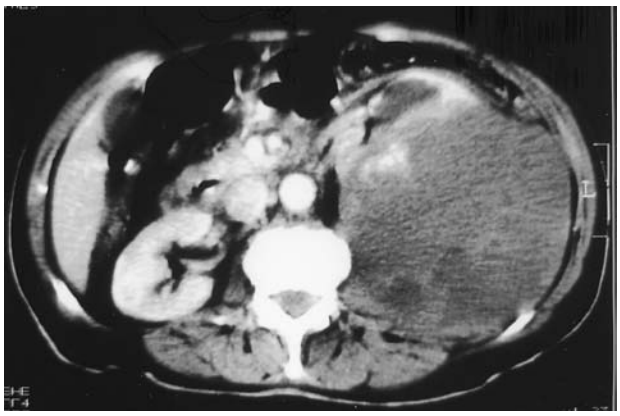

Figure 1 – *Computerized tomography of the abdomen showing a mass in the upper pole of the left kidney with a hypodense central area.*

Patient received blood transfusion with 1200 mL of red blood cells and was submitted to left radical nephrectomy through a transverse laparotomy. Surgical specimen was constituted by the kidney, perirenal fat and the adrenal, measuring 21 x 16 x 9 cm and weighing 1407 grams. In the upper pole of the kidney we have observed an hematoma measuring 14 x 10 x 9 cm that expanded to the perirenal space (Figure-2). After the coagulum was removed we have observed an irregular nut-brown cavity, compromising approximately 20% of the renal parenchyma. Microscopically it was a malignant neoplasm constituted of a number of irregular vascular spaces, anastomosed, covered by discretely pleomorphic cells, with voluminous and hyperchromatic nuclei, with rare stratification and papilliform projections that infiltrated the circumjacent renal parenchyma. Immunohistochemical study showed that neoplasic cells were positive for antibodies anti-CD31, anti-CD34 and anti-factor VIII (Figure-3). Pathologic staging was pT2pN0pMx.

Patient was discharged 11 days after the surgery being, however, re-admitted 15 days after the discharge with emaciation, dehydration, somnolence and mental confusion. Progressed with bilateral bronchopneumonia and death due to sepsis, even though he was submitted to venous antibiotic therapy. No radiotherapy, chemotherapy, crane tomography, bone scintigraphy and necropsy were performed.

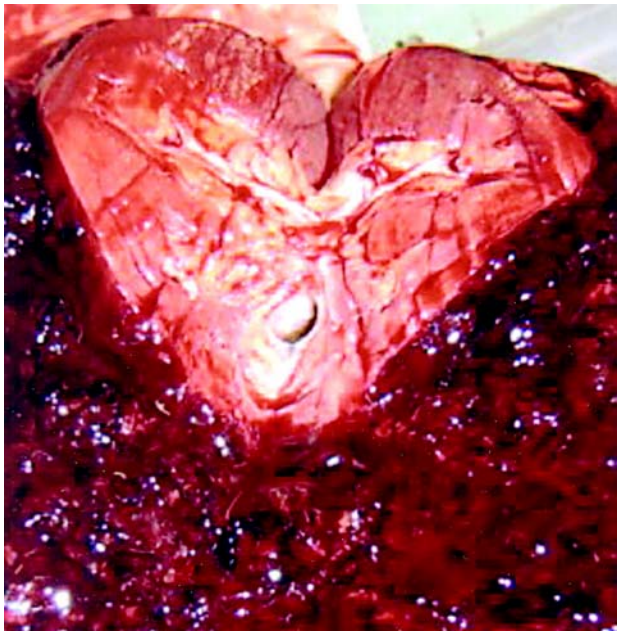

Figure 2 – *Open surgical specimen showing a large hematoma in the upper pole of the kidney, expanded to the perirenal space.*

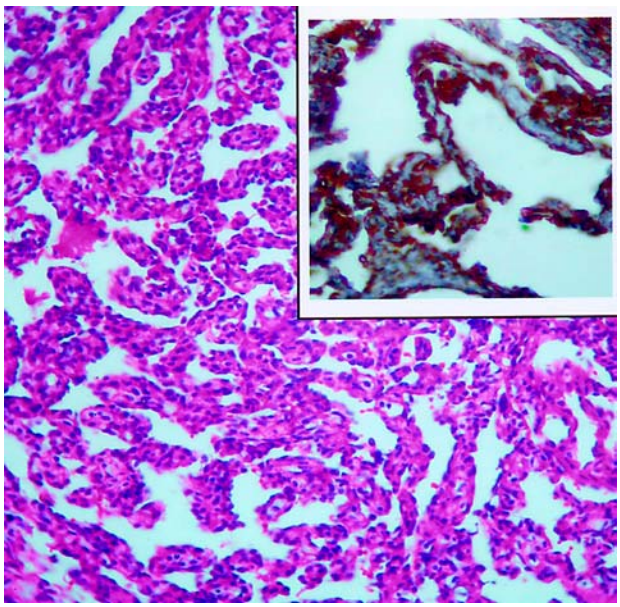

Figure 3 – *Histological section demonstrating an angiosarcoma constituted of irregular vascular spaces, anastomosed, covered by cells with voluminous and hyperchromatic nuclei (HE, X200). Insert - anti-CD31 positive neoplasm cells (Immunostaining, X400).*

COMMENTS

Angiosarcomas are little differentiated neoplasms and, frequently require confirmation by immunohistochemistry. They demonstrate positivity for endothelial cell markers, such as factor VIII, antibodies anti-CD34 and CD31, the latter is more sensible and specific.

These tumors are initially presented in an advanced stage, and liver, lungs and bones metastasis are common. Local recurrence after radical nephrectomy are frequent. Most common clinical manifestations are pain in the flank in 81% of the cases, hematuria in 38% and palpable mass in 31%. Because they are hemorrhagic tumors, patients can usually present anemia. Treatment consists of radical surgery associated to radiotherapy for the local control of the disease. Systemic chemotherapy with cisplatin and isofamide or doxorubicin and isofamide presented a 44 and 71% response rate, respectively. However, patient's mean survival rate is of 13 weeks after diagnosis (2).

Primary renal angiosarcoma should be remembered among differential diagnosis in cases of retroperitoneal hematoma and in hemorrhagic renal tumors.

CONFLICT OF INTEREST

None declared.

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Correspondence address:
Dr. Osmar E. Ribeiro de Souza
Rua Alfen Paixão, 559, bl 4 / 204
Uberaba, MG, 38060-230, Brazil
E-mail: osmaruro@mednet.com.br