

Angiosarcoma of Kidney

A Case Report and Review of Literature

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INTRODUCTION

Angiosarcoma is a rare high-grade malignant tumor accounting for less than 2% of the soft tissue sarcomas.⁽¹⁾ They originate from the endothelium of the blood and lymphatic vessels and can be primary or metastatic, and localized or multicentric. Most angiosarcomas develop in the skin or soft tissue, and others, in the breast, liver, and bones.⁽²⁾ Commonly, metastases occur in the liver, lungs, and bones. Those involving the kidney usually represent metastasis from the skin or the primary visceral lesions. On the contrary, primary angiosarcoma of the kidney is a very rare neoplasm with less than 30 cases reported in the literature to date.^(3,4) To our knowledge, all previous reports described the origin of angiosarcoma in a normal kidney. We report a unique case of primary renal angiosarcoma arising from a multicystic kidney.

CASE REPORT

A 68-year-old man, who was a farmer, presented with a history of mild persistent left flank pain, nocturia, dysuria, and 2 episodes of macroscopic hematuria, starting 1 month earlier. On physical examination, blood pressure was 135/80 mm Hg. Palpation

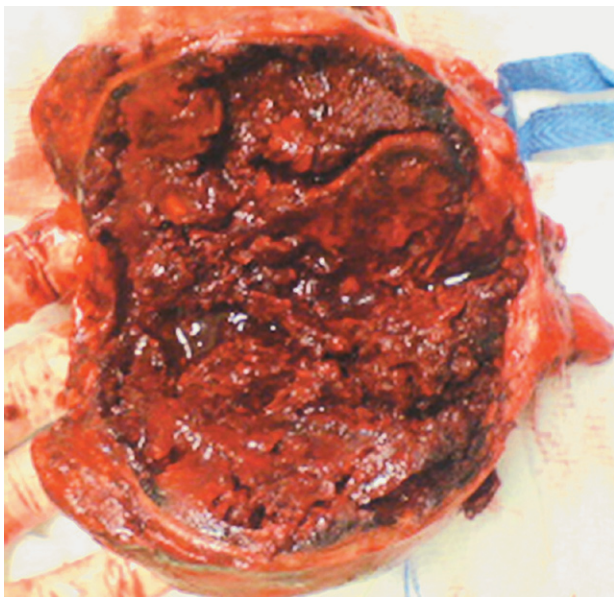
of the lower left quadrant of the abdomen caused diffuse pain and revealed a palpable mass at the left costovertebral angle. On admission, his leukocyte count was $14.5 \times 10^9/L$, 85% of which were granulocytes. Serum levels of sodium and potassium levels were 135 mEq/L and 3.1 mEq/L (reference ranges, 135 mEq/L to 145 mEq/L and 3.5 mEq/L to 5.5 mEq/L), respectively. Kidney function tests showed a blood urea nitrogen of 49 mg/dL (reference range, 17 mg/dL to 53 mg/dL) and a serum creatinine of 1.3 mg/dL (reference range, 0.7 mg/dL to 1.5 mg/dL). Erythrocyte count and hemoglobin concentration were also within normal ranges. Ultrasonography revealed a complex mass of 10 cm in diameter at the normal anatomic position of the left kidney, consisting of cysts with thickened irregular walls and a solid dense central area, suggesting a renal tumor. Abdominal computed tomography with endovenous contrast medium confirmed the above findings. No lymphadenopathy or metastatic diseases were noted. Preoperative diagnosis of cystic renal tumor was made and left nephrectomy was carried out.

The macroscopic appearance of the mass was that of a large solid tumor

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Primary angiosarcoma arising from a multicystic kidney.

mixed with recently clotted blood, replacing four-fifths of the organ (Figure). Diagnosis was made on a morphologic basis, proven by the immunohistochemical study. According to the pathology report, the central area of the mass consisted of irregular vascular spaces, covered by discretely pleomorphic cells with large, hyperchromatic nuclei. Immunohistochemical study showed that malignant cells were positive for anti-CD31 and anti-CD34 antibodies which are specific for angiosarcoma. The patient was discharged 10 days after the surgery and was scheduled for chemotherapy with cisplatin and ifosfamide and short-interval follow-ups, despite the absence of detectable metastases at diagnosis.

DISCUSSION

Predisposing factors for angiosarcoma include kidney transplantation and exposure to arsenic, thorium dioxide, vinyl chloride, and radiation after the treatment of lymphedema, while the causes of primary angiosarcoma in the kidney are still unknown.^(1,5) Occurrence of primary angiosarcoma in a multicystic kidney is intriguing. In fact, this neoplasm is common in both congenital and acquired human renal cystic diseases. An unusual case of mixed Wilms tumor and angiosarcoma in a 38-year-old woman has recently been reported.⁽⁶⁾

Fewer than 30 cases of primary kidney angiosarcoma have been reported to date, and therefore, the natural history of the disease is unknown. The most important prognostic factor is the tumor size and presence or absence of metastases at diagnosis.^(7,8) In tumors larger than 5 cm, the 5-year survival rate is 13%, while this rate is 32% in smaller tumors.⁽⁹⁾ If metastasis is present, survival rates diminish dramatically.⁽¹⁰⁾ Despite the progress in chemotherapy, patients' mean survival rate after diagnosis of metastatic disease remains as low as 13 weeks.⁽¹¹⁾

Local recurrence after radical nephrectomy is frequent in angiosarcomas, since in most of the reported cases, micrometastases had already occurred before diagnosis.⁽³⁾ The usual treatment of localized disease includes wide resection followed by adjuvant radiotherapy, which shows a response rate between 44% and 71%. The role of radiotherapy is controversial.^(12,13) Given the paucity of published cases, it is difficult to standardize treatment of the local disease; it seems however, that the best treatment consists of radical surgery associated with systemic chemotherapy.⁽⁷⁾

Chemotherapy regimens have been used according to the results obtained in other types of sarcomas or in angiosarcomas of other locations. The standard treatment of metastatic disease remains an ifosfamide-based regimen (cisplatin and ifosfamide or doxorubicin and ifosfamide); however, this management provides a median progression-free survival and a median overall survival of about 4 and 8 months, respectively.⁽¹⁴⁾ Taxanes (paclitaxel or docetaxel) may be more efficient in the treatment of metastatic renal angiosarcomas. Paclitaxel has unique activity in angiosarcomas of the face and the scalp; however, its use in angiosarcomas originating from other sites is less well defined. Saroha and colleagues and Skubitz and Haddad showed that paclitaxel was efficient in angiosarcomas originating from other sites (although not statistically superior to nontaxane regimens), while Penel and coworkers reported a 78% nonprogression rate and a 10% complete histological response in patients with unresectable or metastatic angiosarcomas of various origins.⁽¹⁵⁻¹⁷⁾ We still do not know the best

option for patients with metastatic angiosarcoma of the kidney.

Due to nonspecific symptoms and clinical presentation (pain in the flank in 81%, hematuria in 38%, and palpable mass in 31% of the cases), the differential diagnosis is difficult. These tumors are frequently hemorrhagic, and therefore, are able to mimic a retroperitoneal hematoma or cause massive hematuria and anemia.⁽¹⁸⁾ Since they are commonly associated with a renal mass, pathologic examination of the nephrectomy specimen is the only effective method to diagnose angiosarcoma of the kidney.

CONFLICT OF INTEREST

None declared.

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