Clinical Note

Primary angiosarcoma arising from a multicystic kidney

Konstantinos N. Stamatiou, MD, PhD,
Vaios D. Papadimitriou, MD, *Dimitrios M. Takos,* MD,
Ioannis E. Heretis, MD, *Frank A. Sofras,* PhD.

Angiosarcoma is a rare high-grade malignant tumor of primary occurrence that accounts for less than 2% of soft tissue sarcomas.[1] They originate in the endothelium of the blood and lymphatic vessels, and they could be primary or metastatic with various grades of differentiation. They could also be either localized or multicentric. Most of them occur in skin and in soft tissue, while the remaining occurs in viscera such as breast, liver, and bone.[2] Commonly, metastases occur in liver, lungs, and bones. Angiosarcomas involving the kidney usually represent metastasis from skin or visceral primary lesions. On the contrary, angiosarcoma primarily occurring in the kidney is a very rare neoplasm with only 24 cases reported in the international literature up to date.[3] It occurs most frequently during the sixth and seventh decades of life, and with a male predilection.[2] All previous reports described the origin of angiosarcoma in an otherwise, normal kidney. Here, we report a unique case of primary renal angiosarcoma arising from a multicystic kidney. The occurrence of primary angiosarcoma in a multicystic kidney is intriguing and possibly connects its etiopathogenesis with human renal cystic disease. In fact, neoplasia is common to several forms of human renal cystic disease, both congenital, (such as the von Hippel-Lindau disease, the tuberous sclerosis syndrome, and so forth) and acquired, (such as the adult polycystic renal disease). To our knowledge, the etiopathogenesis of this entity remains unknown. Predisposing factors for angiosarcoma occurring, however, in other tissues include: exposure to arsenic, thorium dioxide, vinyl chloride, radiation, and post treatment lymphedema.[1]

A 68 year-old farmer, with a history of mild persistent left flank pain, nocturia, dysuria, and 2 episodes of macroscopic hematuria, starting one month before, presented to the outpatient department of our hospital. On physical examination blood pressure was 135/80. Palpation of the lower left abdomen caused diffuse pain and revealed a palpable mass at the left costovertebral angle. Upon admittance, his white blood cell count was elevated (14500/mm^3 [normal range: 5000-10000/mm^3], granulocytes 85% [normal: up to 60%]), electrolytes were diminished sodium: 135, potassium: 3.1 [normal ranges: 135-145 mEq/lit and 3.5-5.5 mEq/lit), renal function was normal (blood urea nitrogen 49, serum creatinine: 1.3 [normal ranges: 17-53 mg/ml, and 0.7-1.5 mg/100ml]), while red blood cell count and hemoglobin concentration were relatively normal (hematocrit [Hct]: 37.5%, Hb: 12.5mg/dL, RBC: 4.26M/mm^3 [normal ranges: 40-54%, 14-18 g/100ml, and 4.3-5.9 M/mm^3]). Ultrasound revealed a complex mass at the normal anatomic position of the left kidney, that consisted of cysts having thickened irregular walls and a solid dense central area, suggesting a renal tumor. The abdomen computerized tomography with endovenous contrast medium confirmed the above finding. No lymphadenopathy and metastatic disease were noted. Preoperative diagnosis of cystic renal tumor was made, and left nephrectomy was carried out (**Figure 1**). Diagnosis was given on a morphologic basis, and proven by an immunohistochemical study. According to the pathology report, the central area of the mass constituted irregular vascular spaces, covered by discretely pleomorphic cells, with large, hyperchromatic nuclei, with rare stratification. Immunohistochemical study showed that malignant cells were positive for antibodies, anti-CD34, and anti-CD31, which are more sensible and specific for angiosarcoma. He was discharged 10 days after the surgery, and he was scheduled for chemotherapy with cisplatin, isofamide, and short term interval follow up, despite the absence of detectable metastases at diagnosis.

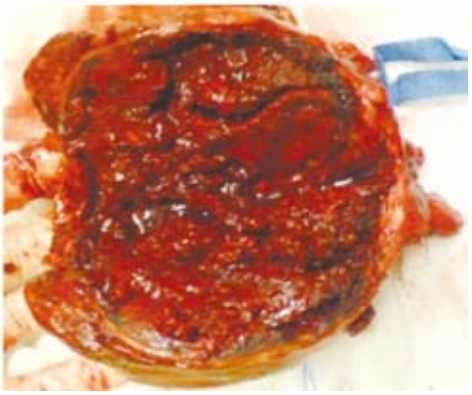

Figure 1 - Primary angiosarcoma arising from a multicystic kidney.

As mentioned above fewer than 25 cases of primary angiosarcoma of the kidney have been reported, and therefore the natural history of the disease is unknown, however, since angiosarcomas are little differentiated neoplasms, in most of the reported cases micrometastases had already occurred at diagnosis, while local recurrence after radical nephrectomy is frequent. For this reason, the patient's mean survival rate is of 13 weeks after diagnosis.[3] Today's datum does not allow us to definitely estimate which treatment represents the best option for patients with endocervicosis of the bladder. However, accordingly to the current experience best treatment consists of radical surgery associated with systemic chemotherapy with cisplatin and isofamide

or doxorubicin and isofamide. Indeed, chemotherapy showed a 44-71% response rate, although durability of the response is likely to be short.[4] On the contrary, radiotherapy for the local control of the disease is controversial. According to some authors, postoperative adjuvant radiotherapy may contribute to local control as it does for sarcomas in other sites, while others stated that radiotherapy does not prolong survival.[5]

Due to non specific symptoms and clinical presentation (pain in the flank in 81% of the cases, hematuria in 38% and palpable mass in 31%), the differential diagnosis is difficult, however, renal angiosarcomas are frequently hemorrhagic tumors, being able to simulate a retroperitoneal hematoma or cause massive hematuria, therefore, they should be suspected in patients presenting with renal mass, hematouria, and anemia. Since it is commonly associated with a renal mass, pathologic examination of the nephrectomy specimen is the only effective method to diagnose angiosarcoma.

Received 15th January 2008. Accepted 28th May 2008.

From the Department of Urology, University of Crete Medical School, Voutes Point, Iraklio, Crete, Greece. Address correspondence and reprint requests to: Dr. Stamatiou N. Konstantinos, 2 Salepoula Street, 18536, Piraeus, Greece E-mail: stamatiouk@yahoo.com

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