

Case Report

Primary Leiomyosarcoma of the Kidney: A Case Report and Review of Literature

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ABSTRACT

Primary sarcomas of the kidney are exceptional. Among renal sarcomas, leiomyosarcoma is the most common histologic subtype amounting to 50-60% of all cases. The tumor usually arises from either the renal capsule or smooth muscle tissue in the renal pelvic wall. In this paper, we report a patient with leiomyosarcoma of the kidney presented with non-specific abdominal pain and a palpable mass. Histologic examination revealed a mesenchymal tumor composed of pleomorphic spindle cells arranged in interlacing pattern with high mitotic activity. Immunohistochemical study was positive for Desmin, smooth muscle Actin (SMA), and muscle specific antigen (MSA). The rarity of this disease and very little information about it, its severity, and poor prognosis, uncommon form of presentation, large size of the mass, and difficulty in management are highlighted and reported.

Keywords: Leiomyosarcoma, Kidney, Iran

Introduction

Primary sarcomas of the kidney are rare. They constitute 1.1% of all malignant renal tumors (1-3) and are seen in only 0.1% of autopsies (1). Leiomyosarcoma, fibrosarcoma and liposarcoma are the most frequent malignant renal neoplasm of pure mesenchymal origin (1, 2), and leiomyosarcoma is the most common type (4, 5). Rhabdomyosarcoma, primary osteosarcoma, chondrosarcoma, malignant neurilemoma, and hemangiopericytoma have also been reported. Clinical manifestations include mass, pain and hematuria. The pain may be dull due to the tumor size, necrosis within the tumor, or the passage of blood clot through the collecting system.

Systemic symptoms such as fever, nausea, anemia, and weight loss are not frequent (1). Here a case of renal leiomyosarcoma in a patient presented with non-specific abdominal pain and a palpable mass is presented.

Case Report

A 56-year-old man presented with a four-month history of right upper quadrant mass and dull abdominal pain, which increased in frequency and intensity during the last two months of this illness. The abdominal pain was later accompanied by mild nausea, heartburn and anorexia. He denied hematuria, frequency, and weight loss. On physical examination,

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the patient was hemodynamically stable and a huge right flank mass was found in palpation. Laboratory findings were within normal limits except for mild microscopic hematuria. CT-Scan of abdomen showed a large heterogenous mass lesion in the right perinephric space with minimal post contrast enhancement and MRI confirmed that the mass originated from the kidney. Right nephrectomy was performed and a lobulaed uncircumscribed non-encapsulated firm huge mass measuring (26×18×18 cm) with multifocal hemorrhage and necrosis arising from the renal hilum was found. Histology revealed a hypercellular neoplastic tissue composed of rather pleomorphic spindle cells arranged in interlacing pattern with high mitotic activity and multifocal necrosis (Fig.1). In addition, focally hemangiopricytoma-like pattern and myxoid appearance were detected. The surgeon designated all specimens as lymph nodes involved by the tumor, but histologically no evidence of lymphoid tissue was seen. Sections were stained for broad panel of immunohistochemical markers, which were positive for Demin (Fig. 2), SMA, MSA and negative for Cytokeratin, S-100, C-Kit and HMB-45. In addition, Ki-67 was positive for 6-10% of tumoral cells.

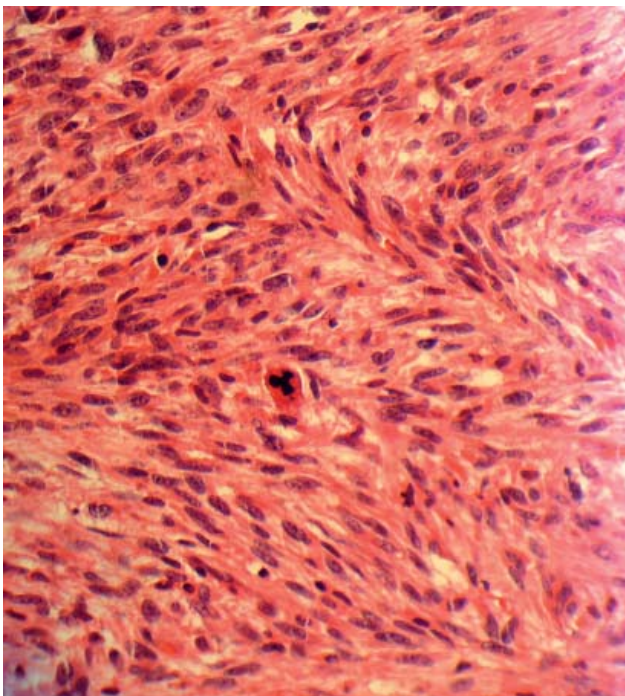


Fig. 1. High power view of tumor revealing pleomorphism of tumoral cells with abnormal mitosis (H&E×400)

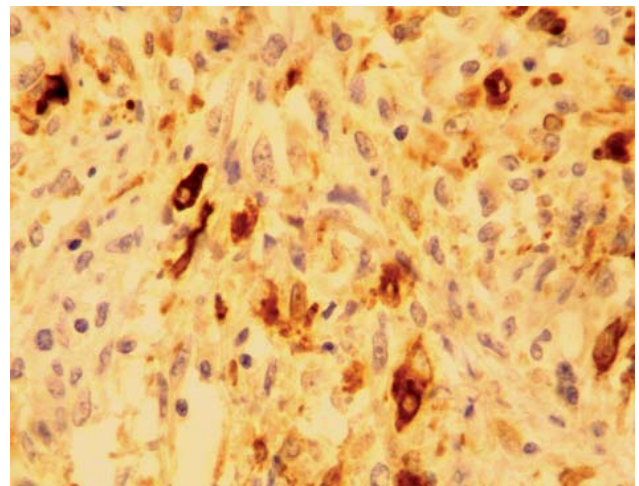


Fig. 2. Immunoreactivity for desmin in neoplastic component

Consequently, histologic diagnosis of leiomyosarcoma was made. The patient was discharged after 10 days while his condition was stable.

Discussion

Sarcoma of the kidney are extremely rare and of varied cell types (2,6). The common signs and symptoms associated with renal sarcoma in adult include a palpable mass, abdominal or flank pain and hematuria, similar to those seen with large, rapidly growing renal cell carcinoma (6). These neoplasms exhibit an aggressive biological behavior and an unfavorable prognosis and are, thus, more lethal than sarcomas of any other genitourinary sites (58-). Renal leiomyosarcoma is the most common histological subtype of renal sarcomas, accounting for approximately 50-60% of the reported cases (4,5,9,10). Men and women are equally afflicted possibly with a slight male preponderance (2, 3). Renal leiomyosarcoma arises from the renal capsule, the smooth muscle fibers of the pelvis or vasculature (5,9,11-) and reportedly capsular leiomyosarcoma is the most common sarcoma of the kidney. Mean age at diagnosis is the sixth decade (4). However, cases in neonatal period have been reported (12). Local recurrence is frequent, particularly if the tumor is not well-circumscribed and has extended beyond the kidney. Lung, liver, and regional lymph nodes are the sites of metastasis (1). There is no pathognomonic finding that would allow differential diagnosis. In the majority of cases, renal leiomyosarcoma is diagnosed through pathologic examination after surgical resection of the renal mass (9, 10).

Spontaneous rupture of this sarcoma is very rare and is commonly seen for large angiomyolipoma, occasional in renal cell carcinoma and Wilms tumor (10). The CT and angiographic findings that may suggest a preoperative diagnosis of renal sarcoma include: (a) A tumor that originates from renal capsule or renal sinus, (b) Lack of extension of the mass beyond its capsule, (c) Fatty density of the mass in cases of liposarcoma (d) and Vascular pattern of the tumor (2). Renal leiomyosarcoma exhibits irregular tortuous tumor vessels without pooling of contrast material or arteriovenous shunting sparse neovascularity (1). Liposarcoma, a common retroperitoneal neoplasm, rarely arises in the kidney and renal lipoma and angiomyolipoma rarely undergo malignant changes (2). Liposarcoma of the kidney often originates in areas that do not contain fatty tissue, such as the renal capsule and, rarely the renal parenchyma. In general, capsular liposarcoma causes inward displacement of the renal parenchyma without renal invasion—a feature that can easily be recognized on CT-Scan (2). Radiologically, it is difficult to differentiate primary capsular liposarcoma from a secondary invasion of the renal capsule by retroperitoneal liposarcoma. However, in the latter group, the tumors generally grow to a very large size and cause significant displacement of intraabdominal organs including kidney (2). Angiography and CT-Scan, illustrating the integrity of the renal capsule, may enable differentiation between these two lesions (12). Liposarcoma has a variety of appearance on CT-Scan that correlated with their histology. For example, two studies showed that capsular location of a fatty tumor that is avascular or hypovascular on angiogram is quite characteristic of renal liposarcoma (1, 2). In contrast, angiomyolipomas are usually hypervascular tumors with small aneurysms, sunburst appearance and without arteriovenous shunting. From all malignant renal tumors, 83% are renal carcinoma. According to Watson (1), 16% are hypovascular and 6% avascular. Therefore, renal carcinoma is that most common etiology for a neoplasm with a hypovascular angiographic pattern. Sarcoma botryoid is a highly malignant neoplasm and may occur in unusual locations where striated muscle is not a normal component. Because rhabdomyosarcoma is frequently found in Wilms tumor, some feel that rhabdomyosarcoma is actually a nephroblastoma in which striated muscle cells are dominant. Angiographic features include moderate neovascularity with arterial encasement, pooling of contrast material and tumor stain.

The treatment of choice is radical nephrectomy (6,7,10), but is rarely feasible due to the invasion of adjacent structures by the tumor (9). The most important prognostic factor is the tumor-free resection margin (13). Large mass size and frequent metastasis to adjacent organs are found in most cases at the time of diagnosis, and, consequently, the prognosis of most leiomyosarcomas is poor (9, 13). Although radiotherapy, chemotherapy, or hormonal treatment is considered as adjuvant treatment, these treatments do not appear to alter the clinical course (14). Analysis of 66 cases of renal leiomyosarcoma by Niceta *et al.* (15) found higher morbidity in females than in males, rapid growth and high metastatic tendency, and both local and distant recurrence of renal leiomyosarcoma. Moreover, almost all patients died within two years despite radical nephrectomy (5).

Miyajima *et al.* (16) suggested that the age and sex of the patient, tumor depth, tumor size, frequency of mitotic figures, degree of necrosis, shape of the nucleus, and AJCC tumor stage are the important prognostic factors. They additionally stated that the tumor size and the AJCC tumor stage are the most important of these prognostic factors. The five-year survival rate for tumors less than 5 cm is 77.44%, whereas the survival rate drops to 41.86% the tumor is larger than 5 cm (16). Janice *et al.* (17) insisted that physicians should be vigilant about taking serial patient histories and performing physical examinations because almost all soft tissue sarcomas recur within the first two years. They suggested that a chest X-ray should be taken every three months during the first two years and thereafter every six months. The CT or MR imaging is useful to evaluate local recurrence and is recommended every six months. Biopsy should be done if any evidence of recurrence is detected. In one experiment, the 5-year survival rate was 29.36% (18).

Primary sarcomas of the kidney are rare conditions. CT-Scan is useful in suggesting the diagnosis of these neoplasms and angiography would be helpful to confirm the diagnosis. If a well-differentiated neoplasm originates from the renal capsule or renal sinus and the tumor is hypovascular or avascular on angiograms, the diagnosis of renal sarcoma should be considered. Leiomyosarcoma even when confined to the kidney have a poor prognosis. Radical surgery offers the best chance of cure. The role of adjuvant chemo-immunotherapy and/or radiotherapy remains debatable due to a paucity of data on the treatment of this rare renal neoplasm and divergent results from

limited experience in the literature.

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