Olgu / Case

Pure primary leiomyosarcoma of the kidney: A case report

Böbreğin pür primer leiomyosarkomu: Olgu sunumu

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Abstract

Pure renal leiomyosarcoma is a rare and aggressive tumor. It is very difficult to distinguish from other renal tumors clinically and radiologically. It resistants to radiotherapy and chemotherapy and treatment is only surgery. Here we presented a case with left renal mass which diagnosed kidney leiomyosarcoma.

Key Words: Kidney, Leiomyosarcoma, Nephrectomy

Özet

Böbreğin pür leiomyosarkomu nadir görülen ve agresif bir tümördür. Renal leiomyosarkomu klinik ve radyolojik olarak diğer böbrek tümörlerinden ayırt etmek oldukça zordur. Renal leiomyosarkom radyoterapi ve kemoterapiye dirençlidir, tedavisi sadece cerrahidir. Burada sol böbrekte kitlesi olan böbrek leiomyosarkomu tanısı alan olguyu sunduk.

Anahtar Kelimeler: Böbrek, Leiomyosarkom, Nefrektomi

Introduction

Leiomyosarcoma is a rare and aggressive smooth muscle cell tumor that can arise from different anatomic sites. They have a tendency to recur locally and metastasis early via the hematogenous route (1). Primary renal leiomyosarcoma presents a diagnostic challenge. It is a rare malignity with only 0.12% of renal tumors confirmed as leiomyosarcoma. There are no trustworthy clinical or radiologic features to distinguish leiomyosarcoma from more common renal malignancies (2). We presented a case with left renal mass which diagnosed kidney leiomyosarcoma.

Case Report

47-years-old male patient who admitted our clinic with left flank pain. Physical examination, urine analysis, complete blood count and blood biochemistry were normal. A computed tomographic scan imaging demonstrated an 7 cm mass from the left kidney; no thrombus in renal vein or cava and neither regional lymph nodes or adrenal gland involved were informed (Figure 1). Open left radical nephrectomy was performed because of left renal mass. Pathological evaluation of the surgical specimen was reported as leiomyosarcoma of the kidney. Tumor was observed at surgical margin. Patient received radiotherapy for positive surgical margin. A metastatic lesion was showed in the liver on computed tomography three months after the operation. Chemotherapy was planned.

Histologically the tumor showed discreate transition from well-differentiated smooth muscle morphology to high-grade pleomorphic morphology with no smooth muscle differentiated (Figure 2). In the well-differentiated areas cells were arranged in fascicular array and interlacing bundles whereas pleomorphic cells were arranged in diffuse sheets. The pleomorphic areas exhibited a number of bizarre atypical cells with numerous multinucleated tumour giant cells. In these areas, there was marked nuclear pleomorphism and hyperchromasia with prominent nucleoli. Areas of necrosis was also noted. Keeping the differential diagnosis of renal leiomyosarcoma versus sarcomatoid variant of renal cell carcinoma, it was decided to employ immunohistochemistry panel comprising of cytokeratin, PAX 8, desmin, h-caldesmon, smooth muscle actin, vimentin and melanocytic markers like HMB

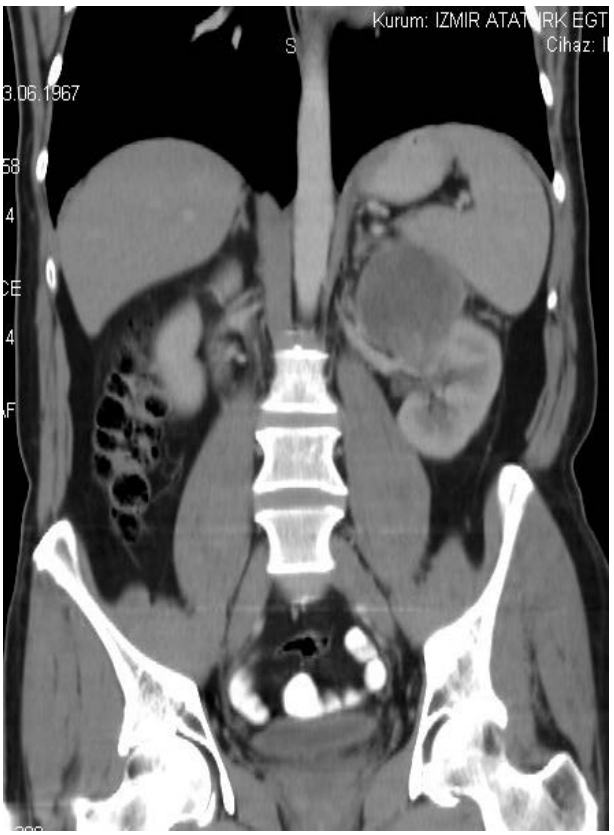

Figure 1. Computed tomographic scan imaging demonstrated an 7 cm mass in the left kidney

45 to rule out other remote possibilities like renal synovial sarcoma and epitheloid angiomyolipoma respectively. Immunohistochemistry revealed strong positivity for smooth muscle actin,desmin, h-caldesmon while all other markers turned out to be negative (Figure 3). Dedifferentiated areas were negative for myogenic markers. This established the diagnosis of renal dedifferentiated leiomyosarcoma.

Discussion

Apart from the uterus, soft tissue leiomyosarcoma commonly occurs in the retroperitoneum, and also arises from the blood vessels. Leiomyosarcomas of nonperitoneal soft tissue sites usually involve the lower extremity but they can occur in the head and neck region also (3]. Primary sarcomas constitute from 0.8 to 2.7% of renal tumors in adults. Renal leiomyosarcomas may arise from the smooth muscle fibers of renal pelvis, renal capsule or renal vessels, last one is the most frequent (4).

Clinical presentation is very similar to more common

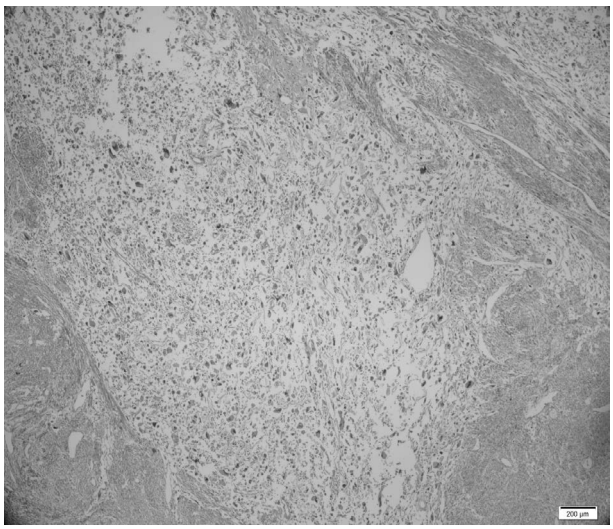

Figure 2. Note the abrupt transition from a differentiated component with smooth muscle morphology to an anaplastic component

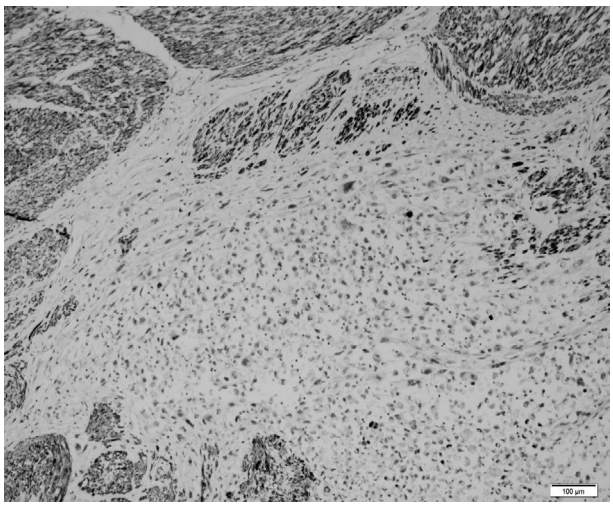

Figure 3. The dedifferentiated component was completely negative for desmin expression (as well as other muscle markers), but classical leiomyosarcoma component was diffuse positive

renal malignancies consisting of flank pain, hematuria, weight loss, and an abdominal mass. Typically occurring between the fourth and eighth decades of life, renal leiomyosarcoma is more common in women and arising from the right kidney (5). It can sometimes present as spontaneous rupture of the kidney (6) Neither ultrasonography, tomography or magnetic resonance are able to differentiate between leiomyosarcomas and renal cell carcinomas (7).

No role for postoperative chemotherapy or radiotherapy has been determinate, although, adjuvant therapy is generally used to tumors with partial resection (8). To date, in complete resection sarcoma, published studies show better local control of the disease but no survival benefit for adjuvant treatment with chemotherapy and radiotherapy (9-11).

There are quite a few cases of primer renal leiomyosarcoma in the literature. Unfortunately it is very difficult to distinguish from other renal tumors clinically and radiologically. Renal leiomyosarcoma resistants to radiotherapy and chemotherapy and surgery is the only treatment option.

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