

Low-grade leiomyosarcoma of renal vein: A case report

Renal venin düşük dereceli leiomyosarkomu: Olgu sunumu

Nalan NEŞE¹, Demet K. ÇAVDAR¹, Bilal GÜMÜŞ², Aydın İŞİSAĞ¹

Celal Bayar University Faculty of Medicine, Department of Pathology¹, Urology², MANİSA

ABSTRACT

Leiomyosarcomas originating from renal vein are quite rare malignant tumors since only 30 cases have been reported in the literature. Clinical symptoms which are due to a renal mass along with radiological findings, usually mislead to a preoperative diagnosis of renal cell carcinoma. The correct diagnosis is frequently made on nephrectomy specimens. Low-grade leiomyosarcomas need to be differentiated from leiomyomas. Increased mitotic activity and necrosis are known to be gold standards for differential diagnosis. In this report, we present a case of a 62 year old woman whose tumor in the left kidney diagnosed as low-grade leiomyosarcoma together with a brief review of the literature.

Key words: Renal vein, leiomyosarcoma, kidney

ÖZET

Renal venden kaynaklanan leiomyosarkomlar oldukça nadir görülen malign tümörlerdir. Bugüne kadar literatürde bildirilmiş 30 kadar olgu vardır. Klinik semptomlar renal kitleye bağlıdır ve genellikle hastalar preoperatif olarak böbrek karsinomu ön tanısıyla opere edilirler. Doğru tanı sıklıkla ameliyat materyallerinde konulur. Düşük dereceli leiomyosarkomlar leiomyomlardan ayrılmalıdır. Artmış mitotik aktivite ve nekroz ayırıcı tanıda önemli kriterlerdir. Sol böbreğinde düşük dereceli leiomyosarkom tanısı alan 62 yaşındaki kadın hasta, klinik ve patolojik özellikleriyle birlikte literatür eşliğinde sunulmaktadır.

Anahtar sözcükler: Renal ven, leiomyosarkom, böbrek

INTRODUCTION

Leiomyosarcomas of vascular origin are uncommon soft tissue tumors and vast majority of them are located in the large vessels such as vena cava and pulmonary arteries (1-4). Leiomyosarcomas quite rarely originate from renal vein (1). In the literature, approximately 30 cases have been reported (2). Preoperative diagnosis is difficult because these tumors are rare tumors and cause nonspecific symptoms (1,2). Low-grade tumors should be differentiated from leiomyomas as seen in our case. We report a case with a low-grade leiomyosarcoma of the left renal vein.

CASE REPORT

The patient was a 62-year-old woman complaining of left flank pain for one month. Ultrasonography and abdominal computed tomography showed presence of a well-demarcated tumor measuring 8.5x6 cm at the hilum of the left kidney. Tumor was nodular, solid and heterogeneous, containing hypodense areas in the center. Preoperative diagnosis was renal cell carcinoma. For treatment, left radical nephrectomy was performed. On gross examination, an encapsulated tumor originating from the wall of renal vein at the hilum of the left kidney (Figure 1a) was observed. Cut surface of the tumor was gray-white, firm and whorled. Central foci of myxoid changes and areas of necrosis were noted (Figure 1b).

Received: 11.11.2007

Accepted: 10.01.2008

Corresponding Author: Dr. Nalan Neşe, Celal Bayar Üniversitesi Tıp Fakültesi Patoloji Anabilim Dalı, Manisa

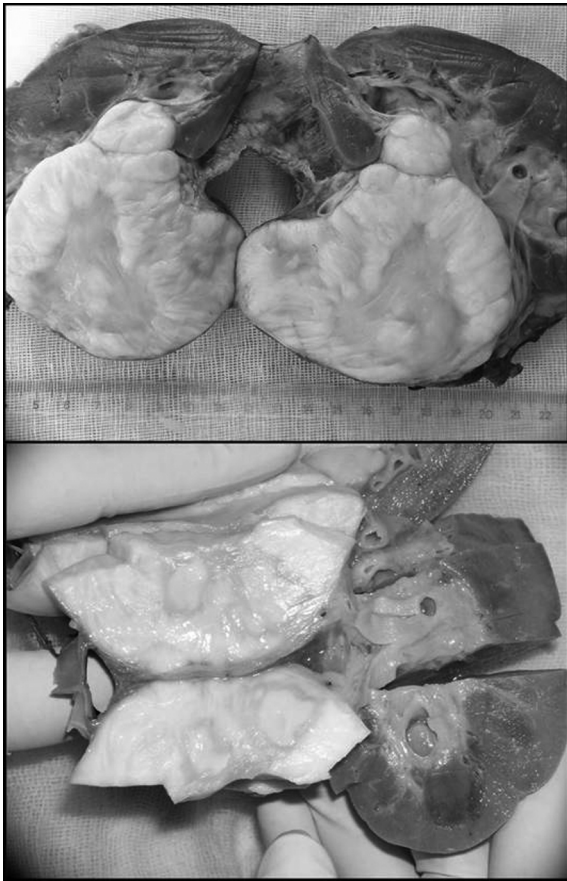


Figure 1. a) Tumor originating from the renal vein in the hilum of the left kidney. Cut surface is gray-white with whorled appearance. Focal myxoid area is seen at the center of the tumor. b) The areas of necrosis are seen.

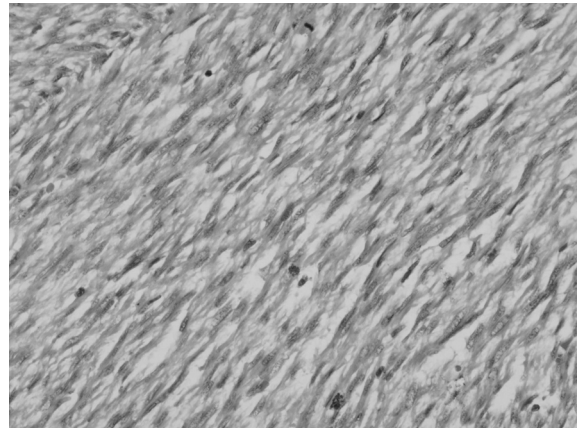


Figure 2. Smooth muscle fascicles made up of spindle-shaped cells with oval tapering nuclei and rare polygonal, large cells with eosinophilic cytoplasm and pleomorphic nuclei are seen. There are atypical mitoses (HE x200).

Microscopical examination revealed typical morphology of leiomyomas in most of the sections, consisting of perpendicularly oriented smooth muscle fascicles made up of spindle-shaped cells with oval tapering nuclei (Figure 2). Rarely polygonal, large cells with eosinophilic cytoplasm and pleomorphic nuclei were seen. Tumor originating from the wall of the renal vein was observed (Figure 3a). Although twenty samples of tumoral tissue were taken for microscopic examination, increased mitotic activity

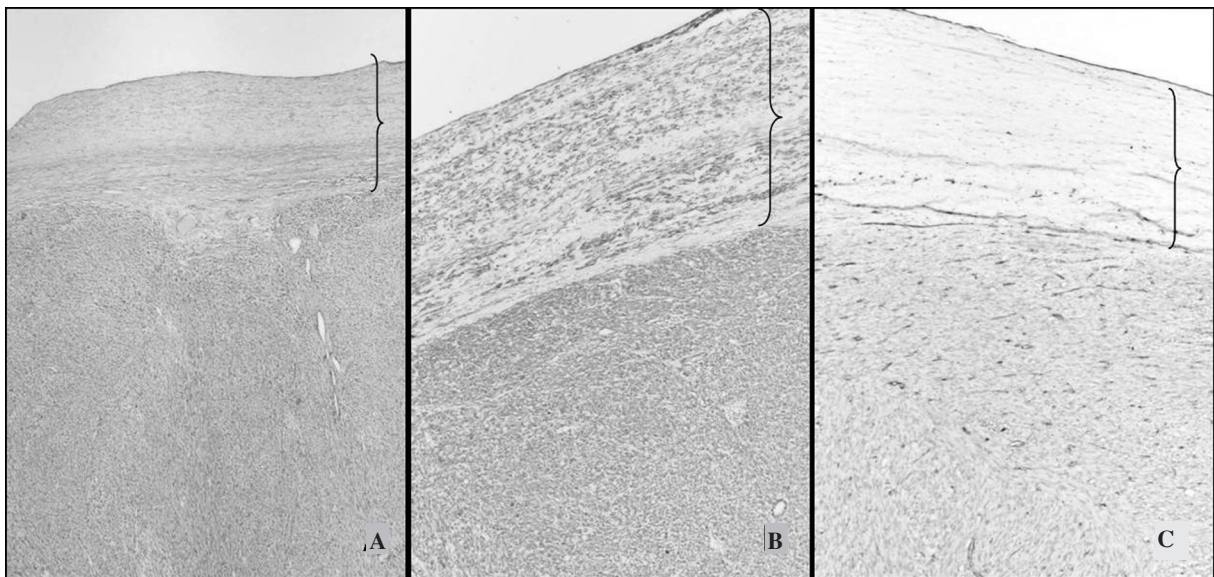


Figure 3. a) Tumor originating from the renal vein (H&E, x100). b) Reactivity with smooth muscle actin dye (DAB, x100). c) Negative staining of the tumor with CD34 while capillary vessels and endothelium of renal vein are serving as positive controls (x100). Renal vein is shown within bracket.

including typical and atypical mitoses were seen in only two sections. Number of mitoses was 6/10 HPF (high power field) in these areas (Figure 2). Focal myxoid change and areas of necrosis were also noted microscopically. No vascular invasion was detected. Immunohistochemically, tumoral cells were stained positively with smooth muscle actin and desmin (Figure 3b). CD34 (Figure 3c) and S-100 protein were not detected. With these findings, tumor was diagnosed as low-grade leiomyosarcoma.

As the tumor was evaluated as low grade and no perirenal infiltration was observed, nephrectomy without chemotherapy or radiotherapy was preferred as a treatment modality. The patient is still alive without metastases or recurrences two years after the diagnosis.

DISCUSSION

Vast majority of leiomyosarcomas originating from vascular structures is usually located in the large vessels such as vena cava and pulmonary arteries (1-4). Renal leiomyosarcomas may arise from renal vein and artery as well as from the smooth muscle cells of renal capsule or muscular wall of renal pelvis (2,5). Leiomyosarcoma originating from renal vein is a quite rare tumor (1). Approximately 30 cases have been reported in the English medical literature up to now (1,2).

Leiomyosarcomas of renal vein are more frequent in women and left kidney is predominantly involved site, as in our case (2,6). This could be explained by growth and proliferation of smooth muscle which could be influenced by estrogenic stimulation (2). Deyrup et al suggested that this predominancy is due to the length of the left renal vein, which is more than the right vein (2).

Preoperative diagnosis for primary leiomyosarcoma of renal vein is very difficult (2). Clinical symptoms which do not differ from other renal tumors include abdominal pain, loss of weight, palpable mass and hematuria (2). Lei-

omyosarcoma can not be differentiated from renal cell carcinoma radiologically (5,7). Diagnosis is classically made by a pathologist as in our case.

Our case represents a low-grade variant which requires differential diagnosis from leiomyoma. The most important tools to make this distinction are mitotic counts and presence of necrosis (2). But while high mitotic activity correlates with a higher rate of metastasis, low number of mitoses does not always indicate a good course (2). It should be emphasized that extensive sampling is required for differential diagnosis of leiomyosarcoma from leiomyoma. Necrosis is seen in 90% of renal leiomyosarcomas and it is considered as a feature of malignancy (6). Myxoid change may be seen focally as in our case, but importance of myxoid change on prognosis is unknown (6). Immunohistochemically, leiomyosarcomas are usually stained with smooth muscle actin and desmin dyes (2,4-6) and occasionally with CD34 (2). The location and the size of the tumor (<3 cm) are the other important prognostic factors of vascular leiomyosarcomas (2,8).

Leiomyosarcomas usually show infiltration into perirenal adipose tissue. Local recurrence is reported in 40% of the cases and distant metastases are primarily to the lungs, followed by liver and bones (1,2,9,10). Metastatic tumors are frequently high grade tumors. No metastases or recurrences were seen in our case during the 2 year follow-up period. Tumors originating from vena cava inferior show more aggressive course than those arising from renal vein (3).

The suggested primary treatment of kidney leiomyosarcoma is radical nephrectomy, followed by chemotherapy and radiotherapy for high grade tumors (1,2). Partial nephrectomy may be another choice of treatment for small sized leiomyosarcomas as Cocuzza et al suggested (9). Large and low-grade tumors are treated by nephrectomy alone as in our case.

REFERENCES

1. Lemos GC, El Hayek OR, Apezato M. Leiomyosarcoma of the renal vein. *Int Braz J Urol* 2003;29:43-14.
2. Aguilar IC, Benavente VA, Pow-Sang MR, Morante CM, Meza L, Destefano V, et al. Leiomyosarcoma of the renal vein: case report and review of the literature. *Urol Oncol* 2005;23:22-26
3. Kwon TW, Sung KB, Cho YP, Kim DK, Yang SM, Ro JY, Kim GE. Pararenal leiomyosarcoma of the inferior vena cava. *J Korean Med Sci* 2003;18:355-359.
4. Brandes SB, Chelsky MJ, Petersen RO, Greenberg RE. Leiomyosarcoma of the renal vein. *J Surg Oncol* 1996;63:195-200.
5. Minami H, Ueki O, Tanaka T, Nishida H, Hashimoto T, Kawaguchi K. Case of leiomyosarcoma of the renal pelvis. *Int J Urol* 2004;11:122-124.
6. Deyrup AT, Montgomery E, Fisher C. Leiomyosarcoma of the Kidney- A Clinicopathologic Study. *Am J Surg Pathol* 2004;28:178-182.
7. Maeda T, Tateishi U, Fujimoto H, Kanai Y, Sugimura K, Arai Y. Leiomyosarcoma of the renal vein: arterial encasement on contrast-enhanced dynamic computed tomography. *Int J Urol* 2006;13:611-612.
8. Soulie M. Leiomyosarcoma of the renal vein. *Prog Urol* 2001;11:502-506 (abstract only).
9. Cocuzza M, Arap S, Lucon AM, Saldanha LB. Renal leiomyosarcoma treated with partial nephrectomy. *Clinics* 2005;60:345-346.
10. Grignon D, Ro J, Papadopoulos N, Ayala A. Leiomyosarcoma of renal vein. *Urology* 1991;38:255-258.