

Kidney Preserving Resection of Leiomyosarcoma with Reconstruction of Left Renal Vein: Case Report

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Abstract

Background: Leiomyosarcoma is a common retroperitoneal sarcomas mostly originating from muscles. Only about 5% of leiomyosarcomas arise directly from large blood vessels and more than 50% of cases originate from inferior vena cava. Primary leiomyosarcomas of renal veins are extremely rare (30 cases). Mostly diagnosed at advanced stages because of nonspecific clinical signs.

Case: A 40-year-old woman was evaluated of a left retroperitoneal mass with severe back pain. CT scan revealed a left retroperitoneal mass 7 cm. in size adjacent to left renal hilum. During the operation; a multilobular, solid tumor in 7 x 6.5 x 3 cm size, surrounding the left renal vein was observed. Further dissection revealed that the tumor was originated from left renal vein wall. Total devascularization and partial left renal vein resection with tumor and end to end anastomosis was performed. Vascular outflow was confirmed with intraoperative and postoperative doppler examination. Pathology was reported as moderately differentiated leiomyosarcoma, originated from renal vein wall with tumor free resection margins and the patient was discharged with adjuvant chemoradiotherapy.

Conclusion: Although the radical nephrectomy is the gold standard approach for malign tumors of the kidney, kidney preserving tumor free resection with vascular reconstruction is a feasible alternative followed by adjuvant chemoradiotherapy and close follow up. Kidney preserving tumor free resection with vascular reconstruction and followed by adjuvant chemoradiotherapy is a feasible alternative instead of radical nephrectomy.

Keywords: Smooth muscle tumors; Leiomyosarcoma; Renal vein

Introduction

Leiomyosarcoma is the third commonest primary retroperitoneal malignancy following liposarcoma and malignant fibrous histiocytoma. However, most of these are intramuscular in origin. Only about 5% of leiomyosarcomas arise directly from large blood vessels [1]. The majority of vascular wall related leiomyosarcomas originate from the inferior vena cava. Primary leiomyosarcomas originating from the renal veins are extremely rare; approximately 30 cases have been documented [2] as almost all being case reports. Most tumors are diagnosed at advanced stages because the disease has a relatively silence course initially and usually presents with nonspecific clinical signs.

Case

A 40-year-old woman presented to our clinic with the complain of back pain. She had no significant medical history. Routine biochemical work-up was normal. Upper abdominal ultrasound revealed a left retroperitoneal mass. The mass was described as a tumor that was not related to renal parenchyma or excretory route but in close contact with the renal vein. Further evaluation with contrast enhanced axial CT scan showed an infiltrative, probably malignant left retroperitoneal mass 7 cm in size neighboring the left renal hilum. After informed consent was obtained, the patient underwent exploratory laparotomy. During the operation, a multinodular solid tumor with a size of 7x6.5x3 cm, originating from the left renal vein was observed (Figure 1). The mass which is strongly adhere was not resected totally without left renal vein. So we decided to do a complete circumferential resection, and reconstruction of the left renal vein. The renal vein was freed from the surrounding tissue along its course preserving left gonadal vein. After en-bloc, kidney preserving resection of the mass including invaded part of the vessel was completed (Figure 2), an end to end anastomosis of the vein was performed using 6/0 polypropylene

(Figure 3). Histopathological examination showed that the tumor was a moderately differentiated leiomyosarcoma originating from the wall of the renal vein. It was also confirmed that all resection margins were negative. The early postoperative period was uneventful and the patient was discharged home on fourth day. Two years after receiving adjuvant chemoradiotherapy she is still doing well without any symptom or sign of recurrence.

Discussion

Primary vascular leiomyosarcomas are quite rare; less than 300 cases have been reported in the literature mostly in the form of single case report. Leiomyosarcoma which is originating from vascular system can be fatal problem. More than half of them stem from IVC with a female predominance of 3:1 [3]. Patients with vascular leiomyosarcoma are usually in their sixth or seventh decade of life, but our case was in her forth decade [4]. The tumor is situated on the left-side 60 percent of the time. As was in the present case, these type of tumors are generally diagnosed at an advanced stage due to nonspecific clinical signs.

As a natural result of extremely rare occurrence of the disease,

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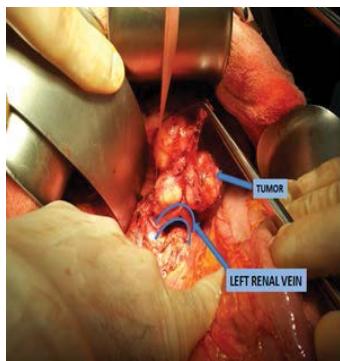


Figure 1: Multilobular mass / Left renal vein



Figure 2: The mass / After resection

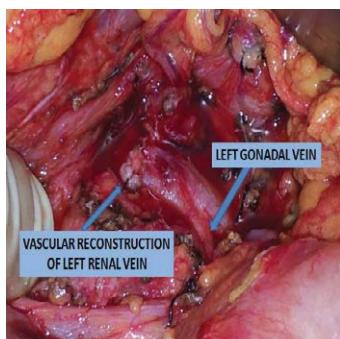


Figure 3: After reconstruction of left renal vein

little is known about the factors impacting on long-term prognosis of patients with leiomyosarcoma of renal vein origin. There is no consensus regarding the optimal treatment strategy [5]. Jihoon T Kim et al. suggested that an aggressive resection with negative margins should be the goal of therapy for those without widespread metastases and who acceptable surgical candidates [6] are.

The most important prognostic indicator for vascular wall originated leiomyosarcoma is surgical resection with negative margins, which confers a 33 to 68 percent 5-year survival [7]. Authors from the University of California reported a five-year survival rate of 53 percent [8]. One of the commonly adopted surgical approaches has been to remove the tumor, en bloc including nephrectomy due to difficult-to-detach adherence to the renal vein [9-11]. We believe that securing a negative vascular margin during resection is adequate and can allow for an organ- or function-preserving removal provided that an appropriate chemoradiotherapy regimen follows when needed. In our case; the tumor was formed in the posterior wall of renal vein. We did not remove the mass, which has tumor free margin, without

circumferential resection of renal vein. Therefore we decided to make a complete circumferential resection and reconstruction of the left renal vein.

To summarize, we believe radical nephrectomy remains to be the gold-standard treatment modality for such malignant tumors; however, R0 resection can be a feasible alternative if supported with adjuvant chemoradiotherapy in those who require it and followed up closely.

Conflict of interest

Authors have no conflict of interest to disclose.

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