



# Leiomyosarcoma of the Right Ovarian Vein: a Case Report with Multimodality Management and Long-Term Follow-Up

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## Introduction

Leiomyosarcoma is a malignant tumor arising from smooth muscle cells in the alimentary tract, retroperitoneum, genitourinary system, or soft tissue. Primary leiomyosarcoma of vascular origin is a rare tumor and comprises less than 2% of all leiomyosarcomas [1]. The inferior vena cava is the most commonly affected vessel followed by the large central veins and then the long saphenous vein [1, 2]. Leiomyosarcomas arising from the ovarian veins are extremely rare. Only a few sporadic cases have been described in previous studies with the development of hepatic metastasis in one case report [2–6]. To the best of our knowledge, this is the first case report in English literature of a retroperitoneal leiomyosarcoma arising from the ovarian vein with interval development of skeletal and adrenal metastases. The radiological images, as well as pathological findings, are illustrated.

## Case Report

A 55-year-old woman presented to our hospital with a complaint of recurrent pain in the right side of the abdomen and flank region for 1 month. Physical examination was negative for a lump or other significant findings. No other symptoms were present. All laboratory findings were normal. Medical and family histories were unremarkable.

Abdominal sonography revealed the presence of a solid heterogeneously hypoechoic retroperitoneal soft tissue lesion

adjacent to inferior vena cava. Contrast-enhanced computed tomography (CT) of the abdomen (Fig. 1a, b) confirmed a 5 × 5.2 cm size lobulated well-defined heterogeneously enhancing mass with areas of necrosis on the right side of retroperitoneum anterior to the kidney and lateral to the inferior vena cava and duodenum. The lesion was seen contiguous with the adjacent vascular structure extending upwards till the right renal vein. No evidence of invasion into adjacent structures was seen. No pre-operative evidence of distant metastasis was seen. A pre-operative diagnosis of a retroperitoneal tumor with possibilities of vascular leiomyosarcoma or extra-adrenal paraganglioma was made.

During surgery, a 5 × 5.5 cm size mobile mass was found to be located in the right side of retroperitoneal space inferior and medial to the kidney. The tumor was arising from the right ovarian vein with proximal dilatation of the vein. The right renal vein and inferior vena cava were not involved. No definite invasion of the adjacent organs including the right kidney, duodenum, and colon was found. Bilateral ovaries were normal. No significantly enlarged lymph nodes were found. The patient underwent radical resection of the retroperitoneal mass including the involved right ovarian vein with more than 5-cm proximal and distal margins.

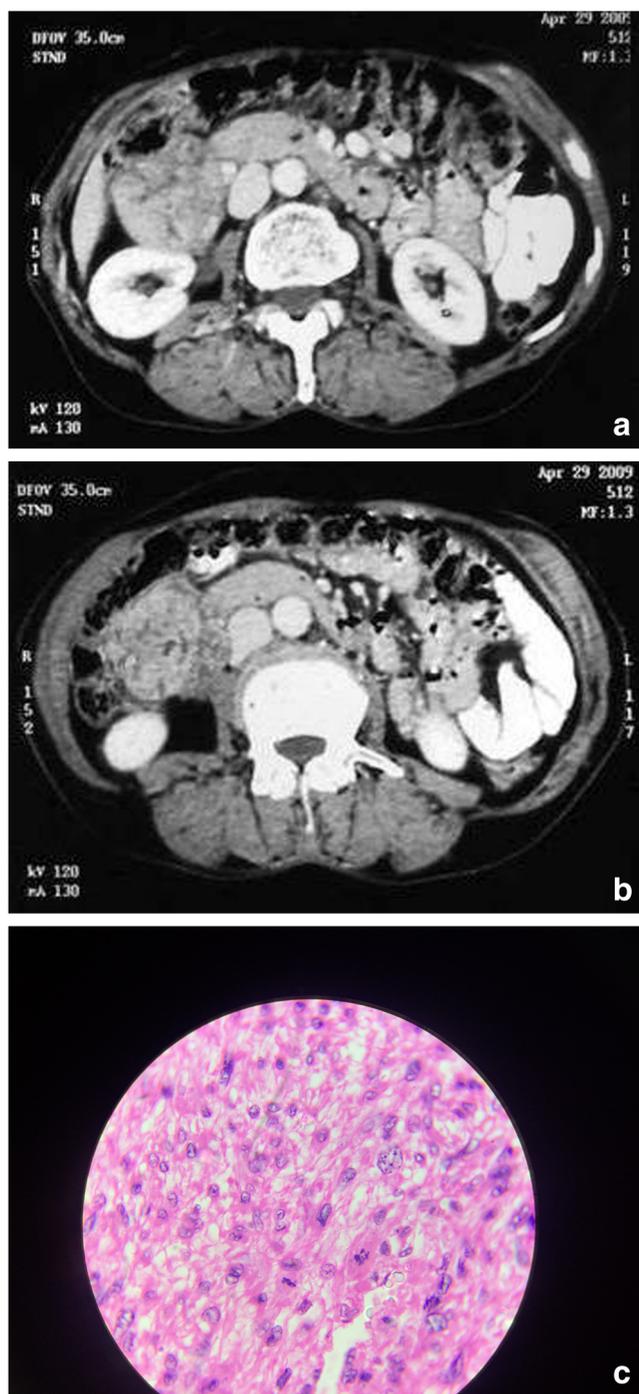
Pathological examination revealed capsulated firm mass measuring 5 × 5 × 4.3 cm showing bosselated and whorled appearance with areas of necrosis on cut section. Histology (Fig. 1c) revealed a spindle cell tumor comprising interlacing bundles of spindle cells with fascicular pattern. The tumor cells were merging with blood vessel walls, palisading nuclei, and eosinophilic fibrillary cytoplasm. Nuclei were cigar-shaped and blunt-ended with variable atypia and occasional multinucleate giant cells. Hemangiopericytoma-like vasculature was noted. Areas of coagulative necrosis were seen. Mitosis was appreciable (10–15/10HPF). Tumor cells were desmin- and SMA-positive and negative for S-100. It was diagnosed as high-grade leiomyosarcoma, with tumor emboli in the ovarian vein.

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**Fig. 1** Contrast-enhanced CT of abdomen (a, b) showing lobulated well-defined heterogeneously enhancing necrotic mass on right side of retroperitoneum contiguous with vascular structure extending upwards till right renal vein (arrow).  $\times 40$  microphotograph of H&E sections of ovarian mass (c) showing fascicles of spindle cells with mild nuclear pleomorphism; the capsule is being lifted by tumor cells

Post-operative period was uneventful. After pathological diagnosis of leiomyosarcoma was made, adjuvant chemotherapy with six cycles of Ifosfamide and

Adriamycin was started. The patient tolerated chemotherapy well and follow-up abdominopelvic computed tomography did not reveal any abnormality.

At 29 months, the patient presented with a new complaint of lower back pain. Magnetic resonance imaging (MRI) of lumbosacral spine (Fig. 2a, b) was performed which revealed the presence of altered signal intensity lytic lesions in the left ala of sacrum, posterior arch elements of the 11th and 12th dorsal (D11 and D12) vertebrae and first and second lumbar (L1 and L2) vertebrae with associated soft tissue component. Nuclear bone scan using  $^{99m}\text{Tc}$ -methylene diphosphonate ( $^{99m}\text{Tc}$ -MDP) demonstrated abnormal uptake in the left sacroiliac region, D11, D12, first to third (L1–L3) and fifth lumbar (L5), and first to fifth sacral (S1–S5) vertebrae suggestive of skeletal metastases. Contrast-enhanced computed tomography (CT) of the abdomen revealed lytic bony lesions in lumbar vertebrae and left ala of sacrum. No lesion was seen in the liver or retroperitoneum. Palliative radiotherapy was initiated at a total dose of 30 Gy delivered in 10 fractions. The patient tolerated radiotherapy well and pain symptoms alleviated.

At 53 months, the patient again presented to our hospital with a complaint of back pain in the thoracic region. MRI of dorsolumbar spine showed the presence multiple altered signal intensity lytic lesions throughout the dorsal, lumbar, and sacral spine. Contrast-enhanced computed tomography (CT) of the abdomen (Fig. 2c) was performed which demonstrated lytic bony lesions and newer appearance of a heterogeneously enhancing soft tissue lesion in the left suprarenal location arising from the left adrenal gland suggestive of metastasis. Palliative radiotherapy was given at a total dose of 20 Gy delivered in 5 fractions from the first dorsal (D1) to ninth (D9) dorsal vertebral region.

## Discussion

Vascular leiomyosarcoma is a rare malignant tumor arising from smooth muscle cells of tunica media of the vessel wall. It accounts for approximately 2% of all leiomyosarcomas and seen five times more commonly in veins than arteries [7]. Retroperitoneal leiomyosarcoma can show completely extravascular (62%), completely intravascular (5%), or both extra- and intravascular (33%) growth patterns [8]. The inferior vena cava is the most common site of origin and accounts for 40–50% of all venous occurrences followed by the central veins and long saphenous vein [1, 9]. Leiomyosarcoma of ovarian vein origin is very rare, only sporadic cases have been reported in the literature.

Retroperitoneal masses are a diverse group and specific diagnosis is difficult to determine due to the overlapping

**Fig. 2** MRI of lumbosacral spine and sacrum (a, b) at 29 months showing skeletal metastases. Contrast-enhanced CT of abdomen (c) at 53 months showing left adrenal gland metastasis



imaging appearances. A retroperitoneal lesion that is separate from retroperitoneal organs and is contiguous with vascular structure should raise the possibility of leiomyosarcoma of vascular origin.

Ultrasonography (USG), excretory urography, computed tomography, magnetic resonance imaging, and venography are the diagnostic modalities used for suspected retroperitoneal lesions. CT is a non-invasive modality of choice for exact localization of mass lesion, demonstration of its relationship with vascular structures, and detection of invasion into adjacent organs as well as distant metastasis. Multiplanar reconstruction (MPR) allows images to be created from the original axial plane in either the coronal, sagittal, or oblique plane, which helps to delineate vascular anatomy and relationship to the mass lesion. In our case, pre-operative CT helped in lowering down the possible diagnosis and assessment of tumor resectability. MRI and nuclear bone scan showed bony metastases and represent the first reported case of leiomyosarcoma of the ovarian vein with bony metastasis imaged by MRI spine and nuclear bone scan.

Complete surgical resection offers the best chance of cure. Leiomyosarcoma remains a potentially lethal disease. The most important prognostic indicator for IVC leiomyosarcoma patients is surgical resection with negative margins, which confers a 33 to 68% 5-year survival rate [10]. Pathology findings relating to tumor grade, mitotic rate, and tumor size have been suggested as possible prognostic factors [11, 12]. Metastasis eventually occurs in most of the survivors of other venous or retroperitoneal sarcomas [1, 2, 8]. Hematological metastasis is more common than the lymphatic spread. Sites

of metastases can be the liver, lung, lymph nodes, retroperitoneum, brain, and bones. Local recurrences also occur. To our knowledge, this is the first case report of leiomyosarcoma of ovarian vein origin with bone and adrenal metastases.

Because of limited experience in the literature, the optimal management of vascular leiomyosarcoma with metastases is unknown. Local excision is offered for retroperitoneal recurrence and metastasectomy for lung recurrence [2, 10, 11]. Adjuvant combination chemotherapy may extend the time to recurrence and overall survival in these patients [13, 14]. In the current study, adjuvant chemotherapy was administered with a combination of Adriamycin and Ifosfamide.

We observed no evidence of local recurrence after surgery. The patient developed bony metastasis after 29 months and was controlled using radiotherapy. The patient again experienced bony metastasis at a different site along with adrenal metastasis after 53 months and received palliative radiotherapy. We believe appropriate adjuvant chemotherapy or radiotherapy can be effective in disease control.

## Conclusion

Primary leiomyosarcomas of ovarian vein origin are aggressive neoplasms and prognosis correlates with the extent of resection, stage, and grade of the tumor. R0 resection offers the best chance of cure. Although metastasis occurred, survival improved with the use of adjuvant chemotherapy and radiotherapy.

## Compliance with Ethical Standards

**Conflict of Interest** The authors declare that they have no conflict of interest.

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