

# Rare Presentation of Retroperitoneal Leiomyosarcoma Mimicking a Myoma in a 46-year-old Woman: A Case Report

**Setareh Akhavan**

Tehran University of Medical Sciences

**Mohadese Peydayesh**

Tehran University of Medical Sciences

**shima alizadeh**

Tehran University of Medical Sciences <https://orcid.org/0000-0001-7882-5633>

**Fatemeh Zamani**

Tehran University of Medical Sciences

**Narges Zamani** (✉ [dr.narges.zamani@gmail.com](mailto:dr.narges.zamani@gmail.com))

Narges Zamani; Department of Oncologic gynecology , Vali-e-Asr Hospital ,Tehran University of Medical Science ,Tehran, Iran

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## Case report

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# Abstract

**Background:** Retroperitoneal sarcoma (RPS) are relatively uncommon, constituting only 10 to 15 percent of all soft tissue sarcomas.

**Case presentation:** We share our experience in encountering RPL with vascular and urethral adhesion in the operation room in a 46-year-old woman, whom was scheduled for surgery as a case of leiomyoma. Huge solid retroperitoneal mass (16\*12\*11 cm) was detected in right broad ligament which was attached to the pelvic floor and seemed to be separated from uterus. Because of a dense adhesion of the tumor to the ureter and vessels tearing of internal iliac vein and ureteral injury occurred during dissection that was repaired. Pathologic examination revealed grade II / III leiomyosarcoma with mitotic rate 12 / 10 HPF, including atypical forms. she candidates for radiation.

**Conclusion:** This case report describes the clinical, imaging, surgical and histopathological findings of retroperitoneal leiomyosarcoma. Due to the rarity of these tumors and the complexity of treatment, evaluation and management should ideally be carried out in a center with multidisciplinary expertise in the treatment of sarcomas.

## Introduction

Leiomyosarcomas, a subset of soft tissue sarcomas, are malignant smooth muscle neoplasms that account for approximately 5–10% of all sarcomas. Natural history depends on the anatomic location in which they arise. About 50% arise in the retroperitoneum/abdomen and these include visceral, uterine, and retroperitoneal, with the uterus being the most common location. Given the frequent enormous size and location of these tumors, abdominal pain, nausea, vomiting, anorexia, weight loss, fatigue, and malaise are commonly presenting symptoms.[1] Retroperitoneal sarcoma (RPS) are relatively uncommon, constituting only 10 to 15 percent of all soft tissue sarcomas.[2] In a population-based series from the Surveillance, Epidemiology, and End Results (SEER) database, the average annual incidence of RPS was approximately 2.7 cases per million population.[3] Leiomyosarcomas of the retroperitoneum arise from the inferior vena cava, its tributaries, or any small vessel. They often present as a mass, and they are usually of an enormous size when diagnosed. To date, most knowledge in treating IVC leiomyosarcoma has come from case reports and case series. Surgical resection offers the best chance at long-term survival and potential for cure. Remarkably, 5-year disease-free survival in patients who underwent resection was between 30% and 60%; however, in those who did not undergo surgery, survival was often less than 1 year.[4–8] We aim to share our experience in encountering RPL with vascular and ureteral adhesion in the operation room on a patient whom was scheduled for surgery as a case of leiomyoma.

## Case Description

A 46-year-old Gravida 3 para 3 woman with no medical, allergy history and drug use was referred to the ward of gynecology and oncology of university-based Hospital with a history of abdominal pain,

hypermenorrhea and pelvic pressure through a few months ago. In vaginal exam there was bulging in the right side which deviated cervix completely to the left and upward position. On abdominal sonography uterus size was 63\*42 mm with an endometrial thickness of 9 mm. A heterogeneous mass of 114\*99 mm size in a posterior cul-de-sac, probably with the uterine origin, was detected. MRI revealed a well-defined heterogeneous soft-tissue mass from degeneration, and necrosis in pelvic cavity posterior to uterus was seen which was separated from uterus and ovaries, in post-contrast images heterogeneous enhancement at the tumor was clear (Fig. 1&2) This mass lesion had pressure effect on right ureter and caused hydronephrosis and was in close contact with right external iliac vein (Fig. 3). Her past surgical history was three times cesarean section with *pfannenstiel incision* and her last child was 10 years old. Attempting for ureteral stent placement before surgery wasn't successful. Hence, she was scheduled for surgery. Fellows of oncogynecology did Surgery under the direct supervision of their attending surgeon. She underwent an exploratory laparotomy under the general anesthesia by a midline incision on suspicion of leiomyoma. Huge solid retroperitoneal mass (16\*12\*11 cm) was detected in right broad ligament which was attached to the pelvic floor. It seemed was separated from uterus. Surgical excision was attempted in order to completely resect the tumor. Because of a dense adhesion of the tumor to the ureter and vessels tearing of internal iliac vein and ureteral injury occurred during dissection. Internal iliac vein was repaired by a vascular surgeon. Intraoperative frozen section analysis suggested smooth muscle tumor without determination of benign or malignant. Hysterectomy and bilateral salpingectomy was done and the ovaries preserved. An internal ureteral DJ stent inserted and primary ureteral repair was done by the urologist. By anesthesiologist consultation we transfused three unit Pack Cell to the patient during operation due to blood loss. Oral diet began one day after surgery and the patient discharged 72 hours after admission. Hb before surgery was 10.5, and she discharged by Hb 9.8. Ureteral DJ removed 6 weeks later with no complication. Pathologic examination revealed grade II / III leiomyosarcoma with mitotic rate 12 / 10 HPF, including atypical forms. Necrosis extent was less than 50% and margin couldn't be assessed. The pathologic stage was pT4. she was a candidate for radiation. External beam radiotherapy was given for 25 sessions.

## Discussion

RPS are relatively uncommon, constituting only 10 to 15 percent of all soft tissue sarcomas.[2] Most tumors are already large and locally advanced at the time they are first detected (median size at diagnosis is approximately 15 cm.[10] Rarely, patients with high-grade, rapidly expanding tumors may experience flu-like symptoms and present with fevers and leukocytosis.[11, 12] Radiographic imaging is a key component of the evaluation of a patient with a retroperitoneal mass. The preferred diagnostic studies are a contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis to evaluate the primary site. MRI with gadolinium is reserved for patients with an allergy to iodinated contrast agents or if there is equivocal muscle, bone involvement on CT. MRI may also be useful for delineating disease in the pelvis. For patients in whom preoperative radiation therapy is being considered, MRI is useful for assessing local tumor extent and surrounding edema, which are optimally included in the treatment volume.[13] In this case, we assumed both sonography and MRI modality. Ultrasound report conducted us

to a large myoma but MRI revealed well-defined heterogeneous soft-tissue mass belike malignant but couldn't definitely specified the origin. The retro peritoneum is the space just posterior to the peritoneal cavity and anterior to the para spinous musculature.[9] The structures of the retro peritoneum include the kidneys, adrenal glands, and perirenal fat bilaterally, the aorta and its major branches (eg, renal arteries), the inferior vena cava and its major tributaries (eg, renal veins), and the bilateral iliac vessels (common, internal, external arteries/veins), and the duodenum and pancreas. In our case there was a mass which originated in this space, probably from an iliac vein with adhesions to ureter. As we all know, the anatomy, there are several vital structures in retroperitoneal space and in our patient because of the history of three cesarean section besides existence of this mass accessing the plane for dissection was abstruse. Determining respectability largely depends on the extension of the tumor and what surrounding structures it involves. A relative contraindication is vascular involvement, because reconstructive options vary given tumor location and tissue involvement.[21]We confronted to the retroperitoneal mass instead of uterus leiomyoma and supposing RPL, the ability to perform a complete surgical resection at the time of initial presentation is the most important prognostic factor for survival.[10, 11, 14–20] Remarkably, 5-year disease-free survival in patients who underwent resection was between 30% and 60%; however, in those who did not undergo surgery, survival was often less than 1 year.[4–8]Our patient was a married 46 years old woman, and we desired complete resection for the better survival. Extensive vascular involvement (aorta, vena cava, and/or iliac vessels), although involvement of the vena cava and iliac veins is a relative, rather than absolute, contraindication for respectability, as these vessels can often be ligated or replaced with interposition grafts.[21] Despite distorted anatomy, we dissected the tumor through adhesions from adjacent structures. The management of bulky intra-abdominal extra-luminal tumors is challenging due to their close proximity or diffusion of other structures. The surgical resection of retroperitoneal leiomyosarcoma (RPLM) can be associated with significant morbidity given that they usually invade important vascular structures such as the inferior vena cava (IVC) and tributaries, the duodenum and the ureter.[22] Management can need to be provided by a specialized team of surgeons. Complete resection often needs extended dissection which may include the vascular structures, kidneys, bladder, and gastrointestinal tract.[23] In our experience we had Urethral Injury that Primary repaired and DJ placement was done with cooperation of the urologist. The most serious incident was tearing off right internal iliac vein through dissection of the mass. Vascular surgeon repaired it. The estimation of blood loss induced us and anesthesiology team for transfusion three units Pack-cell during operation because of blood loss.

Intraoperative frozen section analysis suggested smooth muscle tumor without determination of benign or malignant. Hysterectomy and bilateral salpangectomy was done and the ovaries preserved. She had fine post-operation course without acute complications. The patient was discharged 72 hours after surgery and DJ was removed 6 weeks later without any complication. Permanant pathology report showed leiomyosarcoma but the margin can't be assessed. No pathologic finding was reported on behalf of uterus,cervix and fallopian tubes. Eventually, she candidated for radiation.Van Doorn et al.[24] reported postoperative high-dose radiation therapy in 13 of 34 retroperitoneal sarcoma patients with significantly decreased recurrence rate, nevertheless, the benefits and the effectiveness of radiation therapy have not

been rigorously studied due to scarcity of cases.. Our patient underwent 25 sessions for external radiotherapy. Any discussion on this case would be incomplete without commenting on the ethical aspects of the consent process. she declared that she would be happy the other doctors and medical students learn from the case and didn't mind her condition being discussed. She is currently well 6 months after treatment without recurrence or metastasis.

The COVID-19 pandemic has increased the complexity of cancer care. Important issues include balancing the risk from delaying cancer treatment versus harm from COVID-19, minimizing the number of clinic and hospital visits to reduce exposure mitigating the negative effects of social distancing on delivery of care, and appropriately and fairly allocating limited health care resources. Because of the rarity of these tumors and the complexity of treatment, evaluation and management should ideally be carried out in a center with multidisciplinary expertise in the treatment of sarcomas in a multidisciplinary tumor board.

## Declarations

### **Ethics approval and consent to participate:**

Not applicable

### **Consent for publication:**

Written informed consent was obtained from the patient for anonymized information to be published in this article

### **Availability of data and materials:**

Not applicable

### **Competing interests:**

The authors report no declarations of interest.

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### **Authors'contributions:**

**Akhavan.S:** study concept or design, Investigation performed the surgery **Peydayesh.M.:**, performed the surgery. **Alizadeh.SH:** Writing - Review & Editing; critical revision **Zamani.F:** data collection, Imaging

determination **Zamani.N**: Writing - Review & Editing; critical revision, study concept, performed the surgery

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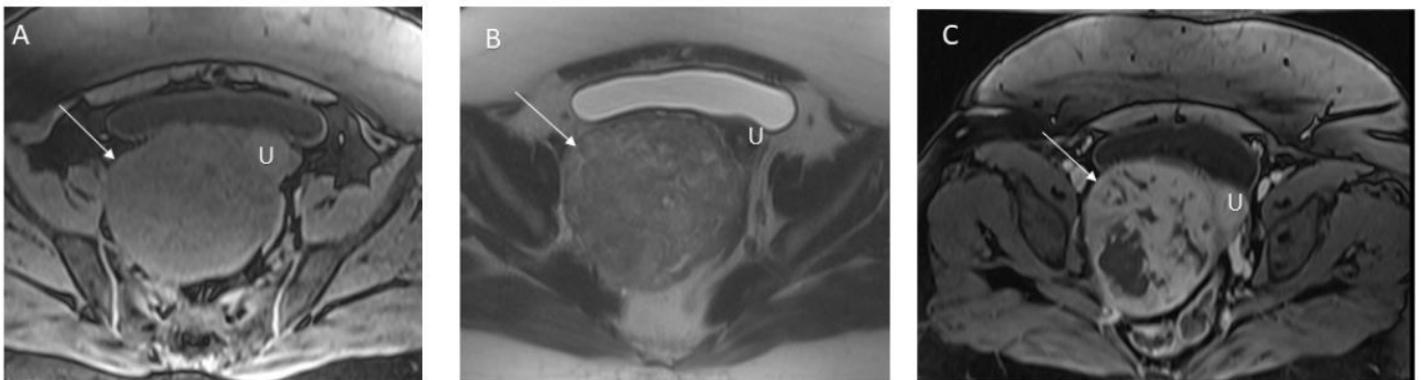
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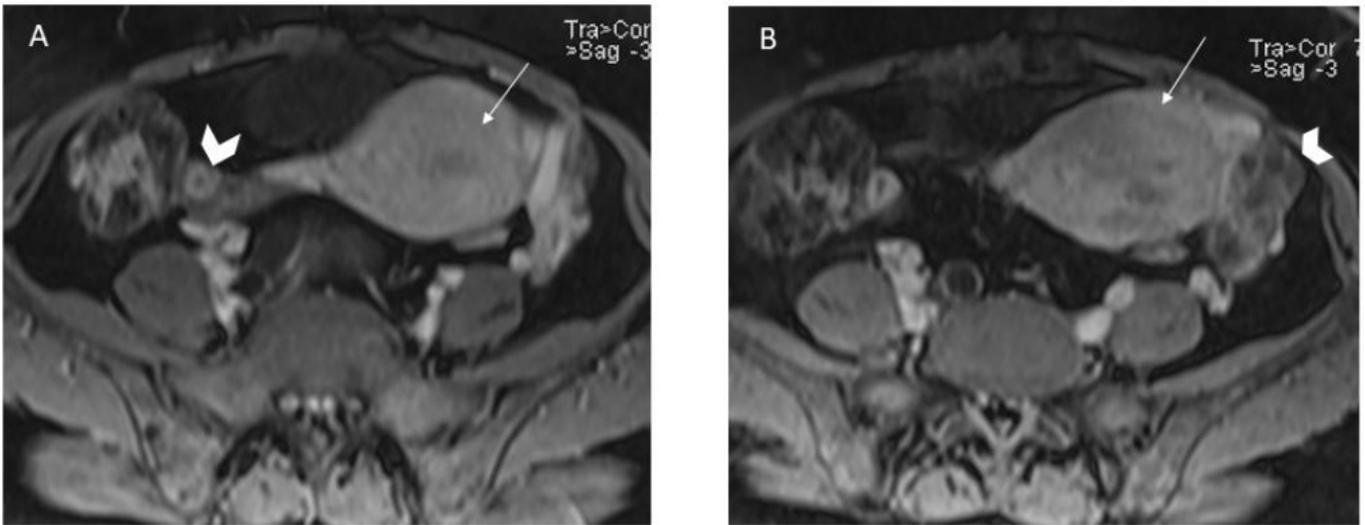
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## Figures



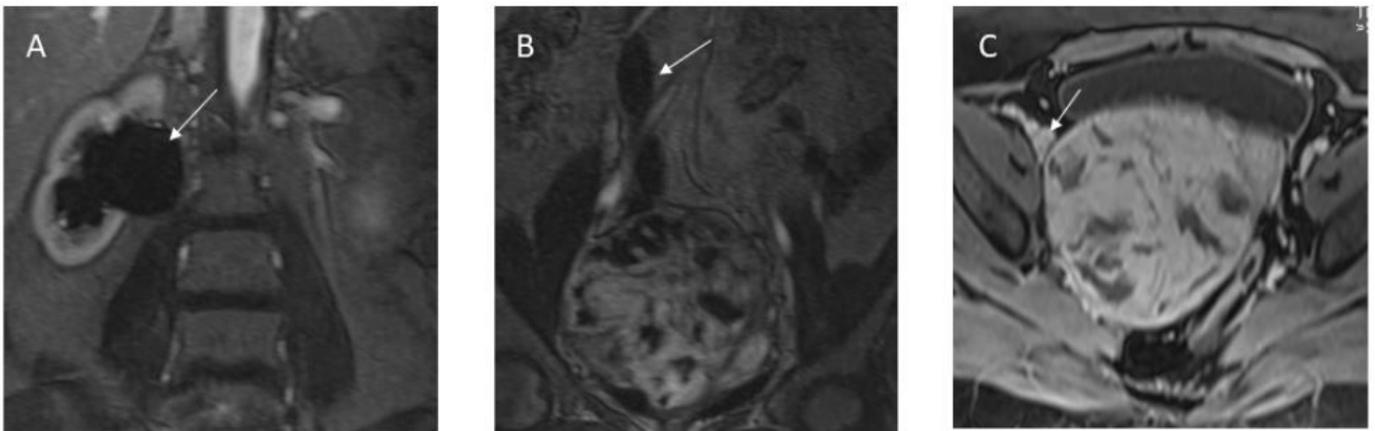
**Figure 1**

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**Figure 2**

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**Figure 3**

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