Retroperitoneal Leiomyosarcoma Invading the Inferior Vena Cava, Uterus and Ovary: Radiologic Findings: Case Report

İnferior Vena Kava, Uterus ve Overi İnvaze Eden Retroperitoneal Leyomiyosarkom: Radyolojik Bulgular

Nilgün IŞIKSALAN ÖZBÜLBÜL, MD,[a]
Muharrem TOLA, MD,[a]
Tülay TEMUÇİN, MD,[b]
Levent IŞIKAY, MD[c]

Departments of [a]Radiology,
[b]Pathology,
[c]Urology,
Türkiye Yüksek İhtisas Hospital, Ankara

Yazışma Adresi/*Correspondence:*
Nilgün IŞIKSALAN ÖZBÜLBÜL, MD
Türkiye Yüksek İhtisas Hospital,
Department of Radiology, Ankara,
TÜRKİYE/TURKEY
nilgunisiksalan@yahoo.com

ABSTRACT Retroperitoneal leiomyosarcoma is a rare neoplasm. Because these lesions are often asymptomatic during growth, the tumor can be massive at the time of diagnosis. About two thirds of leiomyosarcomas are found in the retroperitoneal space. Surgical resection is the treatment of choice, and the most important factor in preventing local recurrence and attaining a good outcome is to be able to remove the tumor completely.; however, complete removal is sometimes difficult if there is an invasion of the surrounding tissues, affecting the inferior vena cava or renal veins. In this case report, imaging findings of a histopathologically proven retroperitoneal leiomyosarcoma in a 38-year-old woman are presented. Also, the benefits of different imaging modalities (Ultrasound, Doppler and MDCT) in the diagnosis are discussed.

Key Words: Retroperitoneal neoplasms, leiomyosarcoma, computed tomography, inferior vena cava, uterus

ÖZET Retroperitoneal leyomiyosarkom nadir bir neoplazmdır. Bu lezyonlar gelişmeleri sırasında sıklıkla asemptomatik oldukları için tümör, tanı anında oldukça büyük olabilir. Leyomiyosarkomların yaklaşık üçte ikisi retroperitoneal boşlukta bulunur. Cerrahi rezeksiyon bir tedavi seçeneğidir ve lokal rekürrensi önlemek ve iyi sonuçlar elde etmede en önemli faktör kitlenin tamamen çıkarılmasını başarmaktır. Bununla birlikte, inferiyor vana kava veya renal venleri etkileyen çevre doku invazyonu mevcut ise kitlenin tamamen çıkarılması bazen güç olabilir. Biz burada, 38 yaşındaki bayan hastada histopatolojik olarak retroperitoneal leyomiyosarkom olduğu kanıtlanmış olgunun görüntüleme bulgularını sunmaktayız. Aynı zamanda, farklı görüntüleme modalitelerinin (ultrasonografi, Doppler ve çok kesitli bilgisayarlı tomografi) faydalarını da tartıştık.

Anahtar Kelimeler: Retroperitoneal neoplazm, leyomiyosarkom, bilgisayarlı tomografi, inferior vena kava, uterus

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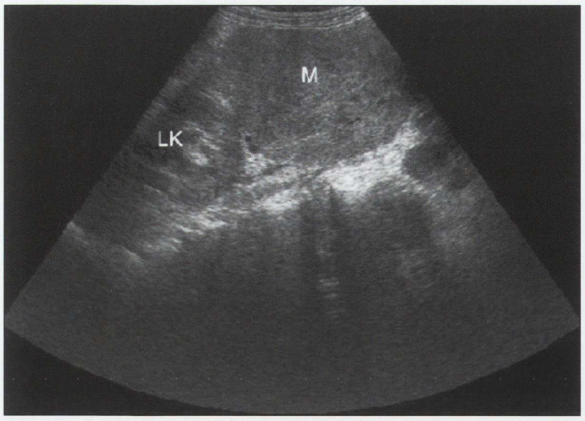
Primary retroperitoneal neoplasms are rare and accounted for only 0.1-0.2% of all malignancies.[1-4] These neoplasms are usually malignant and of mesodermal origin. Leiomyosarcoma (LMS), originating from the retroperitoneum, is further rare and reported incidence being about 11% of all retroperitoneal malignancies.[2] Because these lesions are often asymptomatic during growth, the tumor can be massive at the time of diagnosis.[3-5] Surgical resection is the treatment of choice, however, complete removal is sometimes difficult if there is an invasion of the surrounding tissues affecting the inferior vena cava (IVC) or renal veins.[5] LMS is usually



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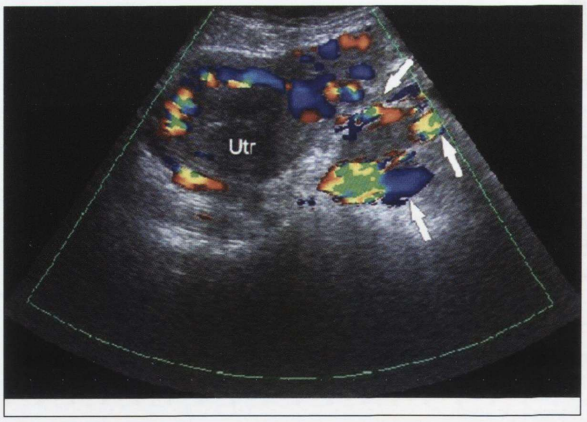

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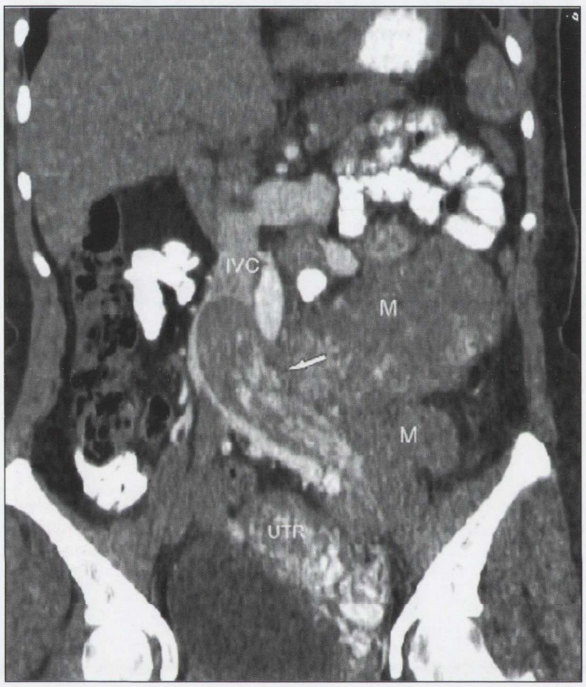

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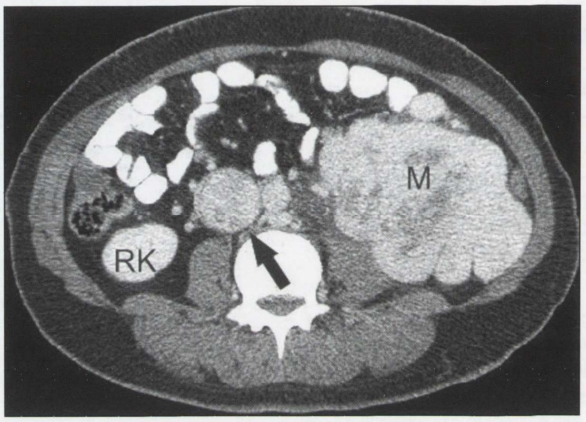

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FIGURE 1: Solid nonnecrotic retroperitoneal leiomyosarcoma in a 38-year-old woman with left flank pain. Sonogram of the left lower quadrant shows a large echogenic mass adjacent to lower pole of the left kidney **(A)**, myometrium is heterogenous and has an increased vascularity **(B)**. Also, note the numerous pelvic collateralls (arrows). Contrast-enhanced CT scan obtained during arterial phase **(C)** shows intense heterogenous enhancement of a mass on sagittal reformatted images. The enhancing tumor thrombus partially occluding and expanding the IVC and the left iliac vein (arrow). Axial contrast-enhanced CT in the portal-venous phase **(D)** demonstrates enhancing tumor and tumor thrombus in the IVC (black arrow).

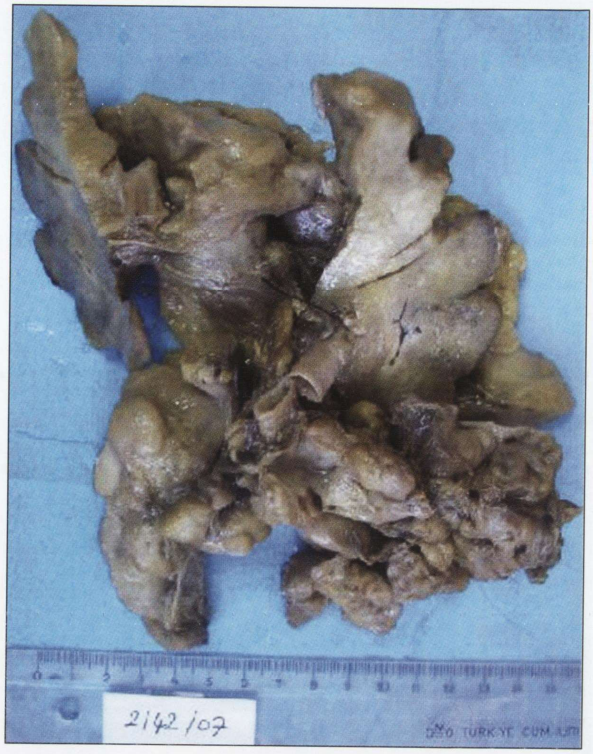
M: Mass, LK: Left Kidney, RK: Right Kidney, Utr: Uterus, IVC: Inferior vena cava.

a well-encapsulated tumor that tends to grow by expansion, rather than invading the surrounding organs.[5] This report describes an unusual case with retroperitoneal LMS invading the IVC, uterus and left ovary at the time of diagnosis. We believed that this is the first report to be describing the imaging findings of visseral and venous invasion in the same patient.

CASE REPORT

38 year-old woman had the complaint about intermittent left flank pain which had started a 2 month ago. Her past medical history was unremarkable. There was no history of vaginal discharge, urinary and bowel complaints. Physical examination was within normal limits. Blood chemistry was normal except for an ESR of 67 mm/hr. Urine analysis showed microscopic hematuria. Abdominal ultrasound revealed the heterogenous echo texture of the solid mass of indeterminate origin in the lower quadrant of abdomen (Figure 1A). Because of the mass extending to the left adnexial loj, transvaginal ultrasound was performed. In this examination, the findings of the invasion of the uterus and left adnexial region such as heterogenous myometrium and non-visualized left ovary seperated from the

mass were detected. The mass was hypervascular on Doppler ultrasound. In addition, there were many collaterals in the left adnexa and low resistance arterial flows in the myometrium (Figure 1B). Subsequently, thin-slice multi-phase (unenhanced, early arterial, late arterial and portal venous phase) computed tomography (CT) was performed to reveal the origin and the extent of the mass and the assessment of the hepatic parenchyma. 16-section multi-detector CT (MDCT) scan demonstrated intense inhomogenous contrast enhancement of the mass on the arterial phase and persisted for late arterial and portal phase. The mass started just below the left kidney in the retroperitoneum and extending to the left adnex. In addition, myometrium showed heterogenous enhancement. The left ovary was not seperately identified from the mass. Peritumoral and pelvic numerous collateral veins were noted. The infrarenal IVC and left iliac vein expanded and contained thrombus which showed heterogenous enhancement during arterial phase suggesting a tumoral thrombus (Figure 1C, D). The bilaterally renal veins were patent. No liver metastases and abdominal lenfadenopathy was detected. At laparotomy, a retroperitoneal mass was partially resected in continuity with the left ovary because the left ovary was invaded by the tumor. No vascular reconstruction was done. The tumor was measured 20 cm in greatest diameter. The cut surface presented a yellow firm tissue with areas of hemorrhage. The histological diagnosis was LMS but the precise site of origin of the sarcoma remained undetermined (Figure 2A, B). The postoperative course was uneventful. No chemotheraphy was given. Nine months later the patient complained of polymenorrhea and menometrorrhagia. Gadolinium-enhanced pelvic magnetic resonace imaging (MRI) revealed multiple residu mass (7 x 3 cm) in the pelvic region and malign thrombus in the IVC. At operation, uterus, the right ovary and partial omentum was removed. The rectosigmoid colon was removed with the tumor and bowel contuinity was restored. At present, 2 years after the first operation, the patient is alive with no symptoms.



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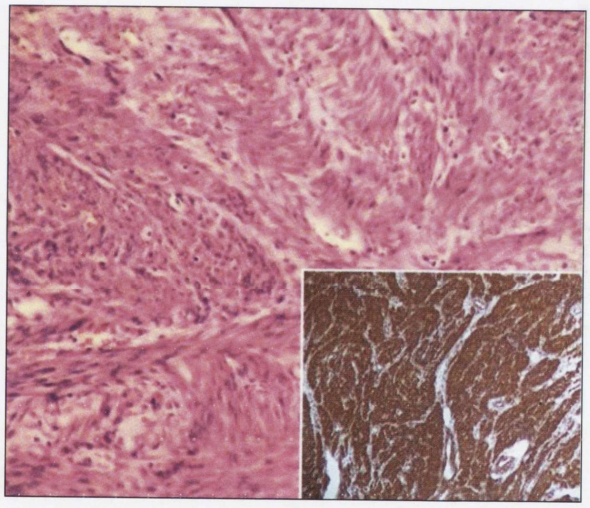

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FIGURE 2: A) Cut section of a retroperitoneal leiomyosarcoma shows well-defined nonnecrotic mass. **B)** Histologic section contains intersecting fascicles with eosinophilic cytoplasm and elongated nuclei with blunt ends (H&E). These cells are immunoreactive for SMA (inset).

DISCUSSION

Retroperitoneal tumors are uncommon, and typically remain aysmptomatic until the tumor becomes evident as a large mass.[3-5] The most common malignant tumors in this site are the lymphomas,

followed by the liposarcomas and other soft tissue sarcomas.[1,3,4] Nonspesific abdominal discomfort or dull pain is a common complaint, caused by the tumor itself and also by compression of the surrounding organs. Other complaints are anorexia, nause and vomiting, back pain, lower extremity swelling, constipation, and weakness.[4-6] In this patient, intermitent left flank pain was the only complaint. Despite uterus and ovaries were invaded, there was no abnormal vaginal bleeding. Her three children were all delivered vaginally at a hospital and home.

Retroperitoneal LMS make-up about half of all soft tissue LMS, and there is a 2:1 female/male ratio, with a median age of 60 years at presentation.[7] A significant proportion of LMS contains large areas of devitalized, necrotic tissue.[1,4,8] Retroperitoneal LMS have a great tendency to develop local recurrences.[5,6] LMS produce metastasis, both lymphatic and hematogenous, earlier in their course than the other retroperitoneal sarcomas.[4,9] In the current case, no distant metastases were detected.

The clinical diagnosis of retroperitoneal tumors is very difficult since the symptoms are extremely variable and nonspecific. The ultrasound and CT findings of a retroperitoneal LMS are not specific and differential diagnosis with other masses can not be made by radiology alone.[6] Ultrasonography shows the consistency of the mass, solid or cystic, and the presence of calcification.[2,6] CT can show the extent of the primary tumor mass, identify metastases, and localize lesions for biopsy.[9] A number of articles have examined the CT characteristics of specific retroperitoneal tumor types such as liposarcomas, leiomyosarcomas and malignant fibrous histiocytomas.[1,3,4,8,9] On CT, leiomyosarcomas are large masses with attenuation equalling to or slightly less than muscle. Occasionally, high attenuation is present due to hemorraghe. Greater than 75% have internal regions of very low attenuation representing necrosis and cystic degeneration. Enhancement is often irregular, mild to moderate in intensity, and predominantly peripheral. Approximately 33% of leiomyosarcomas have both intra- and extravascular components on CT.[1] The ability to make a final histologic diagnosis of a retroperitoneal tumor based on CT characteristics is not often possible, or necessary. Our case showed heterogenous echo texture of myometrium and invasion of the left ovary on ultrasound. Doppler revealed the hypervascularity of the mass and thrombus of the infrarenal IVC. There was no necrosis or cystic degeneration on the mass.

In preoperative planning it is of utmost importance to detect extension of the tumor into the vessel wall as well as other organs.[10] MDCT enables fast and thin acquisition of the abdominal anatomy. This allows multi-phase and multi-planar studies that can be obtained during defined circulatory phases. Multiphase contrast-enhanced CT protocols are recommended for an accurate assessment of the tumor thrombus and its complication. Arterial phase imaging is particularly important if an intravascular lesion is suspected to delineate feeding vessels that may become isodense to the mass on delayed images.[1] Axial images are beter for vessel wall assessment; coronal images provide the best evaluation of proximal tumor extension as well as differentiation between bland and tumor thrombus.[10] In our patient, MDCT with multiplanar reformatting confirmed the extent of the hypervascular mass and invasion of the adjacent organs such as uterus and left ovary in the multi-phase study. Also, tumor thrombus was revealed in the IVC and the iliac vein.

Surgical removal is the treatment of choice as radiotheraphy and chemotheraphy are usually ineffective. More than 70% of patients who underwent surgical resection demonstrated local recurrence.[5] Pathologic examination often fails to determine whether the LMS is originated from the IVC or has invaded the IVC.[5]

In conclusion, retroperitoneal LMS is uncommon. Radiologic findings (ultrasound and CT) are not specific. MDCT can show the tumor extension and the tumoral thrombus on different planes and multiphase enhanced studies. LMS shows a greater tendency to the invasion of the IVC, but at the same time the invasion of the surrounding organs should be checked.

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