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***Case Report** / Olgu Sunumu*

Extrapelvic intravenous uterine leiomyomatosis mimicing cardiac myxoma and deep vein thrombosis

Kardiyak miksoma ve derin ven trombozunu taklit eden ekstrapelvik intravenöz uterin leiomyomatozis

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ABSTRACT

Extrapelvic intravenous uterine leiomyomatosis is a rare smooth muscle neoplasm. Uterine leiomyomatosis is a histologically benign pathology. Rarely, it can be confused with a cardiac mass. A 44-year-old female patient was admitted with increasing severity of pain and swelling in both legs for the past week. The patient was initially diagnosed with bilateral deep vein thrombosis. After further evaluation, we decided that the patient had cardiac myxoma. However, we intraoperatively observed that the lesion in the right atrium was arising from the inferior vena cava. In the final postoperative histopathological evaluation, the definite diagnosis was extrapelvic intravenous leiomyomatosis. The patient was discharged uneventfully following her second operation.

Keywords: Intracardiac leiomyomatosis/leiomyoma, intravenous leiomyomatosis/leiomyoma, myxoma.

ÖZ

Ekstrapelvik intravenöz uterin leiomyomatozis nadir görülen bir düz kas neoplazmıdır. Uterin leiomyomatozis histolojik olarak iyi huylu bir patolojidir. Nadiren, kardiyak kitle ile karıştırılabilirler. Kırk dört yaşında kadın hasta son bir haftadır her iki bacağında giderek artan ağrı ve şişlik şikayeti ile başvurdu. Hastaya ilk olarak bilateral derin ven trombozu tanısı konuldu. Daha ileri değerlendirmeler neticesinde kardiyak miksoma olduğu sonucuna varıldı. Ancak ameliyatımız sırasında kalbin sağ atriyumundaki kitle lezyonunun inferior vena kavaya doğru uzandığı gözlendi. Ameliyat sonrası son histopatolojik değerlendirmede kesin tanı ekstrapelvik intravenöz leiomyomatozis idi. Hasta, ikinci ameliyatı sonrasında sorunsuz şekilde taburcu edildi.

Anahtar sözcükler: İntrakardiyak leiomyomatozis/leiomyoma, intravenöz leiomyomatozis/leiomyom, miksoma.

Extrapelvic intravenous uterine leiomyomatosis (IVUL) is a rare smooth muscle neoplasm. The source of the pathological problem is typically in the uterus, and although uterine leiomyomatosis is histologically benign, it may show clinically malignant behavior by intravascular spread to the adjacent lymphatic or venous system.[1] According to a recent analysis, a total of 110 cases of intravenous leiomyomatosis in 32 different manuscripts reported worldwide were found to have spread to the cardiac area.[2] In these

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studies, approximately 30% of the patients were preoperatively diagnosed with right atrial myxoma. Obstruction in the venous system during the course of the disease may be detected as deep vein thrombosis (DVT) and may be misleading, as was in our case.[3,4]

In our article, we aimed to share our clinical and two-stage surgical experience in a patient who was discharged with bilateral DVT treatment in his first admission and was mistakenly diagnosed with myxoma, which was found to be IVUL after two different operations.

CASE REPORT

A 44-year-old female patient was admitted to the cardiovascular surgery outpatient clinic with increasing severity of pain and swelling in both legs for the past week. Laboratory values for protein S and antithrombin-III activity were 37.7% and was 1.8%, respectively. The genetic thrombophilia panel was negative. In the bilateral venous evaluation of the patient, no thrombosis was found in the main femoral vein and distally. In the abdominal ultrasonography, echogenicities that was presumed to be compatible with the thrombus material were observed within the inferior vena cava (IVC). The patient was discharged when her complaints regressed with anticoagulant treatment (standard heparin in the first three days and then unfractionated heparin + warfarin), and the appropriate international normalized ratio value was achieved. A month later, the patient was readmitted to our clinic with a prediagnosis of myxoma as a result of transthoracic echocardiographic (TTE) examination performed in another medical center where she was admitted due to syncope. The diagnosis of the patient was confirmed by TTE and cardiac magnetic resonance imaging (MRI) in our hospital.

Transesophageal echocardiography (TEE) reported a right atrium giant multilobular mass lesion moving to the right ventricle with a transition from the patent foramen ovale, which prolapses at every systole, to the left atrium, and a myxoid image in favor of a thrombus extending from the IVC (Figure 1a). Magnetic resonance imaging revealed a right atrial mass in favor of myxoma and that the lesion in the IVC might be in the form of a thrombus (Figure 1b).

An intraoperative TEE was placed for the cardiac operation. Extracorporeal circulation was obtained via the right femoral vein and vena cava superior (VCS). The cardiac arrest site was reached by antegrade cardioplegia. The right atrium was opened, and it was directly observed that the tumor was free in the right atrial space. The mass originated from the IVC (Figure 2a). Macroscopically, the tumor was a yellowish-white, elastic, smooth, mobile, polypoidal mass and resistant to manual pulling (Figure 2b). The mass excision was achieved as much as possible by pulling from the IVC towards the cardiac chambers, and the lesion was sent for histopathological examination after the operation. The mass was successfully removed, and a further stepwise abdominal operation was decided. Following bleeding control maneuvers, the patient was decannulated.

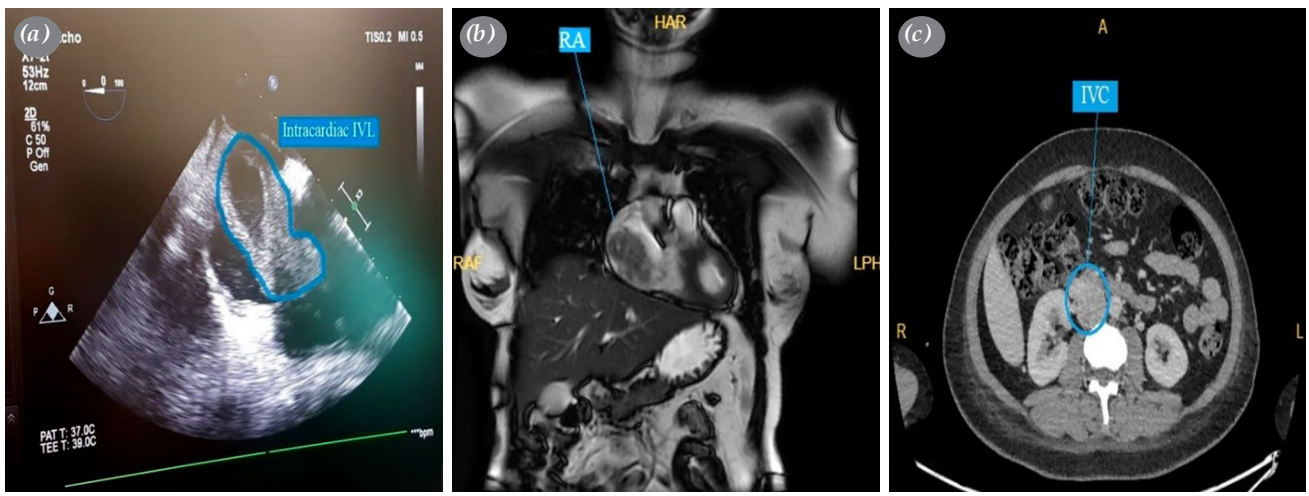

Figure 1. (a) Mobile IVUL between the right atrium and the right ventricle by preoparative TEE, **(b)** Tumor visible in the right atrium on cardiac MRI before planned myxoma operation, **(c)** The IVC filled with tumor mass lesion in contrast-enhanced tomography scan of the abdomen after cardiac surgery.

IVUL: Intravenous uterine leiomyomatosis; TEE: Transesophageal echocardiography; MRI: Magnetic resonance imaging; IVC: Inferior vena cava

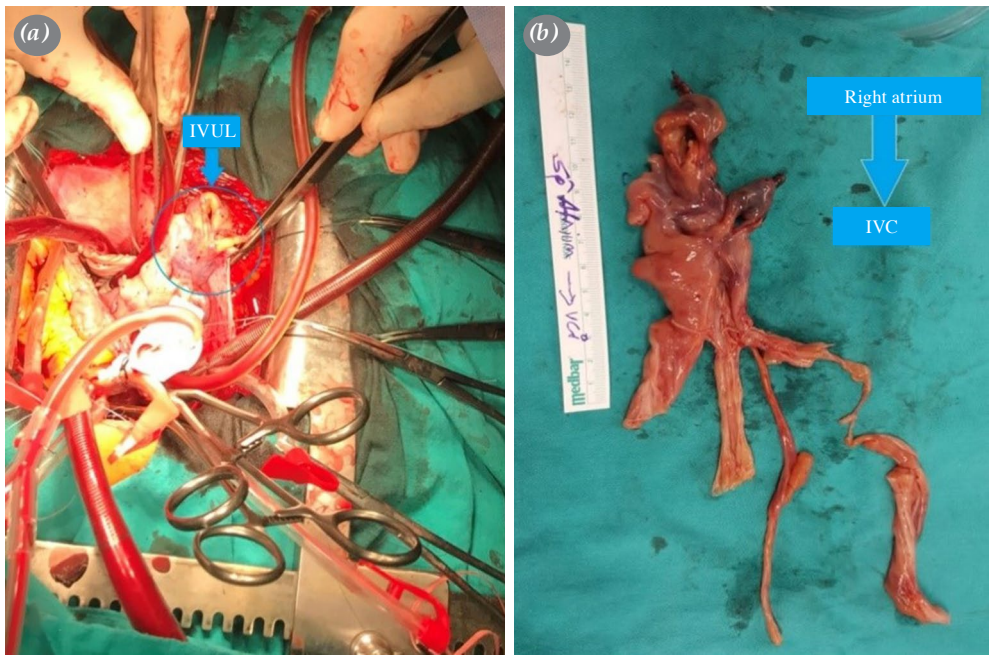

Figure 2. (a) The image of the IVUL in right atriotomy, **(b)** IVUL that is understood to extend from the right atrium to the IVC, excised by pulling as much as it can be delivered.
IVUL: Intravenous uterine leiomyomatosis; IVC: Inferior vena cava.

The extracorporeal circulation circuit was closed. The operation was terminated uneventfully. The patient was transferred to the cardiovascular surgery intensive care unit. Histopathological diagnosis was reported as leiomyomatosis. The postoperative period was uneventful. In the thoracoabdominal computed tomography (CT) after the cardiac mass removal operation, it was reported that the tumor of gynecological origin suggested spread through the venous system (Figure 1c). For the differential diagnosis, uterine leiomyomatosis was diagnosed by whole abdominal MRI.

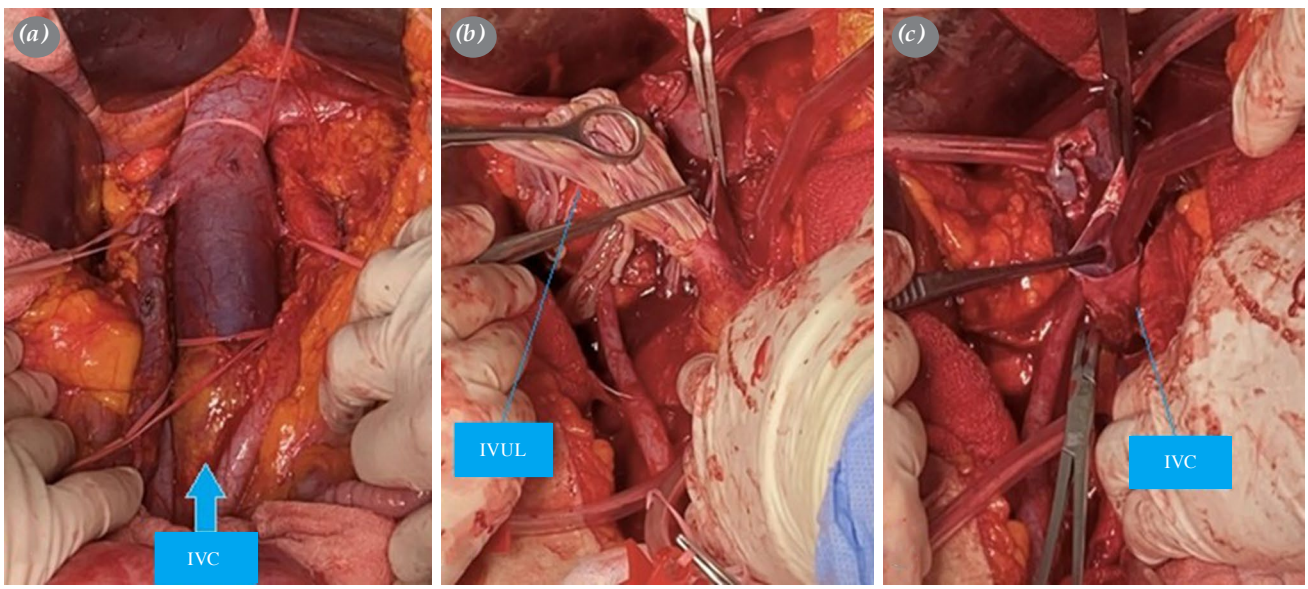

Figure 3. (a) Initial caval visualization, **(b)** IVUL excision, **(c)** post-venotomy.
IVC: Inferior vena cava; IVUL: Intravenous uterine leiomyomatosis.

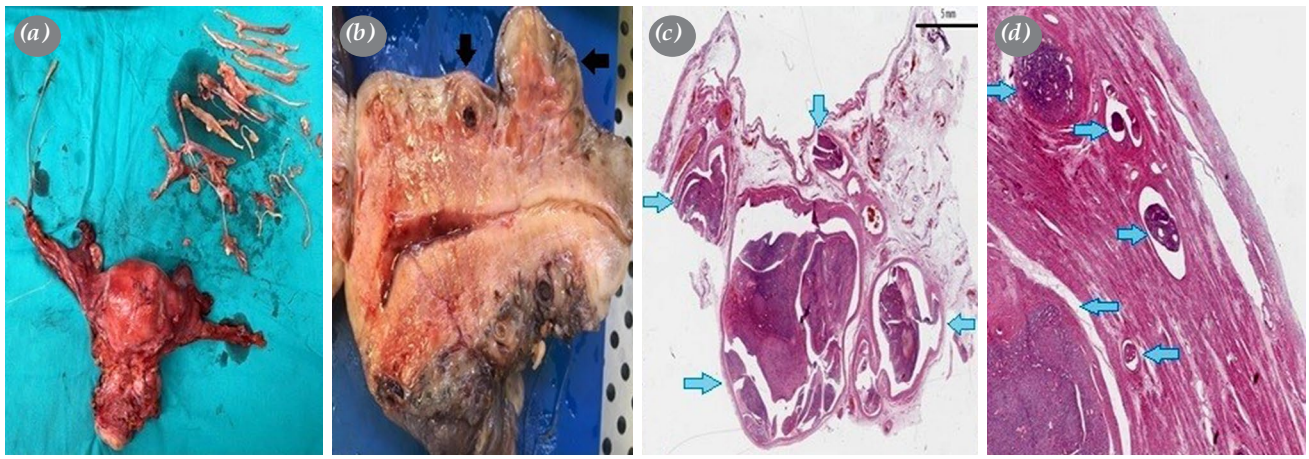

Figure 4. (a) Macroscopic view of hysterectomy specimen and leiomyomatosis materials, **(b)** Leiomyomatosis foci (arrows) in the paracervical region of the horizontal hysterectomy section, **(c)** Leiomyomatosis foci (arrows) within the periadnexal vascular structures (hematoxylin and eosin staining), **(d)** Leiomyomatosis foci (arrows) within the vascular structures in the uterine myometrium (hematoxylin and eosin staining).

Approximately two months after the first operation, the patient was rehospitalized for the complete resection of the pelvic tumor extending into IVC. The patient was operated on together with oncological gynecologists. First, laparotomy was performed, and then total abdominal hysterectomy and bilateral salpingoophorectomy were performed (Figure 4a). Following hysterectomy, the IVC and iliac veins were visualized. Iliac veins were turned with vascular tapes. Vascular structures were prepared for clamping and venotomy. We applied cross clamps to the iliac veins. At this point of surgery, venotomy was performed, and vein chambers were opened (Figure 3). The tumor mass at the proximal end extending to the heart sprang out of the incision by itself with blood flow. Venotomy was also applied to the external iliac vein apart from the IVC; thus, we were able to reach three different vascular lumens. Both main iliac veins and external iliac veins luminal masses were removed. After this stage, bleeding control was cautiously performed. The site was flushed with the appropriate saline fluids. The opened vascular structures were primarily sutured. Distal clamps were removed to provide deairing outflow. The venous intraluminal flow was restored. Surgical drains were placed. Materials were sent for histopathological evaluation. The laparotomy was routinely closed. The patient was discharged two weeks after surgery. Postoperatively, all tumor samples were fixed with standard formalin and divided into multiple blocks (Figure 4b). Pathological sections were stained with hematoxylin and eosin and the IVUL diagnosis was confirmed (Figures 4c, d).

DISCUSSION

Most cases tend to originate from Asia, but there is no clear data that the disease is more common in Asia due to its rarity. At present, the mechanism of IVUL is still not understood. It is most commonly encountered in the fifth decade. The clinical course and symptoms of IVUL often depend on the extent of the tumor. It usually reaches the heart when symptoms begin. The tumor spreads to large caliber vessels, heart, and pulmonary arteries, causing cardiac symptoms, such as cardiac murmur, fainting, pulmonary embolism, and sudden death. The most common clinical complaints are orthopnea, dyspnea, leg edema, arrhythmias, palpitations, heart failure, and syncope.[2]

The first clinical presentation in our patient was swelling and pain in both legs. However, when the second admission was syncope, TTE was to be planned. Due to the insidious course of the disease, early diagnosis is difficult but important for prognosis.[2] Detection and diagnosis modalities include TTE, CT, and MRI.[2] Since advanced CT and MRI provide much more reliable anatomical information, they can minimize misdiagnosis.[5] Unfortunately, the correct differential diagnosis could not be made in our patient as preoperative abdominal imaging was limited to abdominal ultrasonography. Transthoracic echocardiographic plays an important role in the detection of an intracardiac mass. Unfortunately, it may be insufficient according to our experience. We recommend advanced imaging of the abdomen, especially in the presence of DVT and an intracardiac

mass. In addition, we believe that TEE, which was also used in the second operation, provides more accurate intraoperative images in the evaluation of a right atrial mass.

Histopathological examination of our patient at the first operation revealed proliferating smooth muscle fibers without abnormal mitotic activity. Tumor cells were positive for smooth muscle actin and desmin.

There is no consensus on the optimal surgical strategy. Overall, surgical strategies for IVUL vary. Complete resection with total hysterectomy and bilateral oophorectomy is recommended. Recurrence in the first five years is reported between 25 and 33% in patients who do not receive complete resection.[6] Incomplete resection is the cause of postoperative recurrence and long-term death. Ligation of the source vessel is mandatory to prevent recurrence. In our case, the bilateral internal iliac veins and the gonadal vein were meticulously ligated. Since postoperative recurrence of IVUL may occur after surgery, long-term follow-up is required and annual CT examination is recommended.

Routine anticoagulation therapy is required due to the high risk of thrombotic complications in the perioperative period. In the literature, it is recommended to extend anticoagulation therapy up to three months after surgery.[7] The oral anticoagulant drug was discontinued and unfractionated heparin was started since our patient, who received anticoagulant treatment for DVT, had a planned operation, and anticoagulation with unfractionated heparin was continued until the 10th day after the catheter removal since a urethral double-J stent was placed due to postoperative urine leakage. Depending on the risk factors in the preoperative coagulation panel, a new generation oral anticoagulant treatment was started in the outpatient clinical follow-ups.

In conclusion, our experience from this clinical case and our review of the medical literature indicates that intravenous uterine leiomyomatosis can occur with an onset of deep vein thrombosis or cardiac mass lesion. Preoperatively, these cardiac mass lesions may misleadingly present a false myxoma diagnosis. Intravenous uterine leiomyomatosis is a complicated surgical condition that should come to mind in cardiac mass lesions showing inferior vena cava extensions. In our opinion, an intravenous uterine leiomyomatosis diagnosis requires surgery without delay, and surgery necessitates a multidisciplinary approach.

Patient Consent for Publication: A written informed consent was obtained from the patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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