CASE REPORT

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Intravenous Leiomyomatosis with Intracardiac Extension: An Extremely Rare Case

Esra KELEŞ[a], Emre MAT[a], Hakan HANÇER[b], Didem CANOĞLU[c], Ayşen GENÇOGLU[c], Gazi YILDIZ[d], Rezzan Berna TEMOÇİN[d], Pınar BİROL İLTER[d], Emine TULUHAN[d], Medine KAHRAMAN KAYA[d], Ahmet KALE[d]

[a]Department of Gynecologic Oncology, University of Health Sciences Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye
[b]Department of Cardiovascular Surgery, University of Health Sciences Kartal Koşuyolu High Specialization Training and Research Hospital, İstanbul, Türkiye
[c]Department of Pathology, University of Health Sciences Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye
[d]Department of Obstetrics and Gynecology, University of Health Sciences Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye

ABSTRACT Intravenous leiomyomatosis (IVL) with intracardiac extension is a rare disease. It is characterized by the proliferation of uterine smooth muscle cells through the inferior vena cava (IVC) and the right heart chambers. A 36-year-old woman with a history of previous myomectomy, presented with high blood pressure. A transthoracic echocardiography revealed a mass in the IVC. Magnetic resonance imaging demonstrated a large intravascular mass extending from the pelvis to the right heart chambers. The tumor was completely removed in concomitant cardiac surgery and laparotomy. The postoperative course was uneventful. Six months later, the patient was feeling well and in good clinical condition. The histological analysis was compatible with IVL. Intracardiac leiomyomatosis is a rare clinical condition. Multimodality imaging can be helpful in the preoperative diagnosis, although the final diagnosis is based on histopathological assessment. Complete removal of the tumor is curative and has a favorable long-term outcome.

Keywords: Intracardiac; intravenous leiomyomatosis; uterine leiomyoma

Intravenous leiomyomatosis (IVL) is a rare form of uterine leiomyoma, originating from leiomyoma or smooth muscle of uterine vessels.[1,2] It exhibits the characteristics of a benign tumor but behaves like a malignant tumor due to intravascular invasion.

Early diagnosis is difficult because the disease is rare and does not show symptoms until the tumor extends into the heart chambers.[3] The main treatment for patients with IVL is surgery, often requiring a one-step or two-step surgical approach.[4]

Here, we present a case of intracardiac IVL who was successfully treated with a two-step surgical approach. In addition, our report summarized the important clinical characteristics, histopathological features, surgical data, and follow-up results of this rare tumor.

CASE REPORT

A 36-year-old, G3P2A1, multigravid woman presented to the cardiology department for complaining of a headache. She denied chest pain, dyspnea, or palpitations. She had no comorbidities. Her past surgical surgery was significant for cesarean sections and a myomectomy. Additional workups revealed the presence of high blood pressure (140/90 mmHg). Transthoracic echocardiography performed for the evaluation of hypertension inciden-

Correspondence: Esra KELEŞ
Department of Gynecologic Oncology, University of Health Sciences Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye
E-mail: dresrakeles@hotmail.com

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tally identified a mass in the inferior vena cava (IVC) in the right atrium with a diameter of 4.8x2.4 cm that was initially interpreted as metastasis or thrombus. Bilateral carotid and vertebral artery Doppler ultrasonography were normal. Because of these findings, a comprehensive evaluation was carried out for possible underlying diseases. Magnetic resonance imaging revealed a heterogeneous abdominopelvic mass reaching to the right atrium from the level of the right external iliac vein (3.2x3.6 cm, Figure 1). A multidisciplinary examination revealed that this was IVL arising from uterine myoma extending into gonadal vein and penetrating into the heart. Under the double setups, total hysterectomy with bilateral salpingo-oophorectomy was performed, and then the right gonadal vein was resected (Figure 2). Histopathology analysis was consistent with intravenous leiomyomatosis (Figure 3). Second, cardiac surgery was performed after one month. The postoperative course was uneventful, and the patient was discharged on day 5. Six months later the patient was feeling well and in good clinical condition.

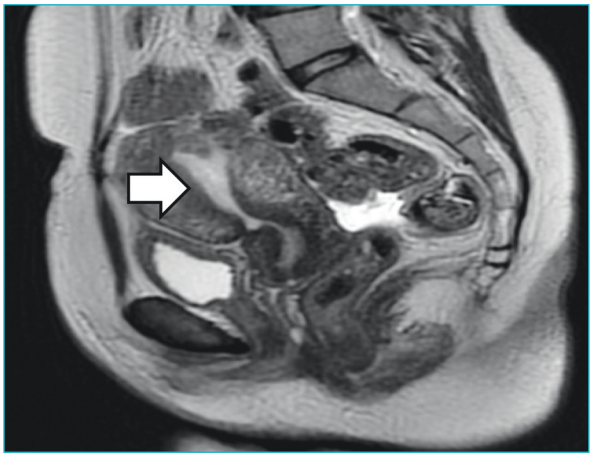

FIGURE 1: Sagittal plane of the pelvic magnetic resonance images showed a heterogeneous abdominopelvic mass starting from the level of the right external iliac vein and extending to the right atrium (arrow).

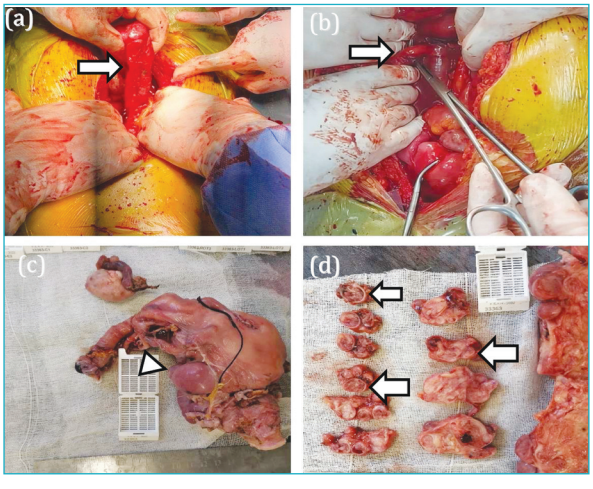

FIGURE 2: a) The right ovarian vein was expanded and full of tumor (arrow); b) Arrow refers to the right ovarian vein (arrow); c) Macroscopic examination showed a large worm-like mass extending the right parametrial wall of uterus (arrowhead); d) Serial sections revealed an extensive intravascular plug filling the vascular lumens (arrow).

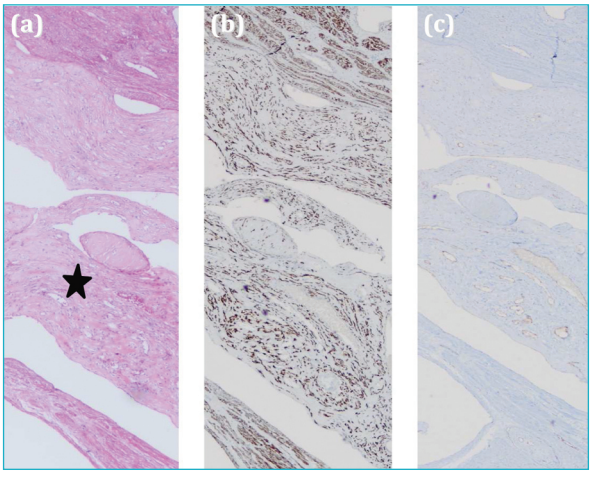

FIGURE 3: **a)** Intravascular tumor almost completely filling the lumen of a vein with uniform spindle-shaped smooth muscle cell proliferation with hyalinized stroma (Hematoxylin and eosin (H&E), magnification x40). **b)** Immunohistochemical staining for Desmin, a smooth muscle marker, showing a staining of intravascular leiomyoma and muscular wall of the vein (Immunohistochemical staining for Desmin, magnification x40). **c)** Immunohistochemial staining for CD31, an endothelial marker, showing staining an endothelial covering of the intravascular leiomyoma (Immunohistochemial staining for CD31, magnification x40).

The patient has given informed consent for this case report.

DISCUSSION

IVL is a distinct type of benign smooth-muscle cell tumor that forms in the uterus and spreads within venous channels.[5] Although histologically benign, IVL has the potential to metastasize to the IVC in approximately 10% of cases and to the right atrium and ventricle in 3% of cases.[6] It affects approximately 85% of premenopausal women with a history of hys-

terectomy or myomectomy, or with concomitant uterine leiomyoma.[7] We describe a premenopausal woman affected by IVL with a right atrial mass with a history of myomectomy.

Prior studies have reported that IVL occurs in women aged 23 to 80 years, mostly with symptoms such as chest pain, vaginal bleeding, palpitation, shortness of breath, dyspnea, and lower leg edema.[8] A 36-year-old woman with a complaint of high blood pressure was presented in this report.

Total surgical excision of the tumor is the primary treatment modality in patients with IVL. The treatment can be performed as removal of the intrathoracic and the abdominopelvic tumors in 2 separate surgeries, or removal of both tumors in a single operation.[9] Prior studies proposed that the two-step approach is safer and easier.[10] We, therefore, opted for a two-staged approach. Since the growth and development of the tumor appear to be estrogen-dependent, it is necessary to remove both ovaries. The presence of estrogen and progesterone receptors was positive in our case.

Since IVL is a very rare condition, it is usually discovered after an operation. It was possible to make a diagnosis in the preoperative period when the patient exhibited no cardiac symptoms and was in good health.

Early diagnosis is often difficult due to the rarity of the pathology, subtle clinical manifestations, cardiac involvement in advanced stages, and poor awareness. However, clinicians should keep IVL in mind in a premenopausal woman with a history of leiomyoma, the presence of cardiological symptoms, and evidence of an intra-cardiac mass in the right atrium. Surgery is the best method of treatment.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız;* ***Design:*** *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız;* ***Control/Supervision:*** *Esra Keleş, Emre Mat;* ***Data Collection and/or Processing:*** *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız, Rezzan Berna Temoçin, Pınar Birol İlter, Emine Tuluhan, Medine Kahraman Kaya, Ahmet Kale;* ***Analysis and/or Interpretation:*** *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız, Rezzan Berna Temoçin, Pınar Birol İlter, Emine Tuluhan, Medine Kahraman Kaya, Ahmet Kale;* ***Literature Review:*** *Esra Keleş, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız;* ***Writing the Article:*** *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız, Rezzan Berna Temoçin, Pınar Birol İlter, Emine Tuluhan, Medine Kahraman Kaya, Ahmet Kale;* ***Critical Review:*** *Esra Keleş, Emre Mat, Hakan Hançer, Didem Canoğlu, Ayşen Gençoğlu, Gazi Yıldız, Rezzan Berna Temoçin, Pınar Birol İlter, Emine Tuluhan, Medine Kahraman Kaya, Ahmet Kale;* ***Materials:*** *Rezzan Berna Temoçin, Pınar Birol İlter, Emine Tuluhan, Medine Kahraman Kaya.*

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