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Intravenous Uterine Leiomyomatosis with Inferior Vena Cava and Intracardiac Extensions[1]

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Intravenous leiomyomatosis originating from the uterus and involving the right ventricle, right atrium, inferior vena cava and iliac veins is an uncommon disease. We report here on a leiomyomatosis in a 35-year-old woman who presented with tumors in her heart and inferior vena cava. The correct diagnosis was achieved by using the transthoracic echocardiographic, cardiac CT and pelvic MRI findings.

Index words : Uterine neoplasms
Leiomyoma
Heart
Ultrasound (US)
Tomography, X-ray

Intravenous leiomyomatosis is a rare benign neoplasm that affects women (1). This tumor often co-exists with uterine leiomyoma or leiomyosarcoma and it represents a cardiac extension through the lumen of veins (2, 3). Here, we report on a case of uterine leiomyomatosis, which presented with tumor progression through the right iliac vein, along the entire inferior vena cava (IVC) and up to the right atrium and ventricle.

Case Report

A 35-year-old woman was admitted to our hospital with a recent history dyspnea upon effort. The patient had undergone uterine myomectomy 10 months previously. The physical examination revealed a heart rate of 86 beats/min with sinus rhythm and a blood pressure of 130/80. There was no heart murmur. The blood counts and blood chemistry were within the normal ranges. The peak serum levels of the creatine kinase-MB fraction (CK-MB) and troponin T were 4.2 ng/mL (normal range: 0–5.8 ng/mL) and 0.06 ng/mL (normal range: 0–0.1 pg/mL), respectively. The level of pro-BNP (B-type natriuretic peptide) was elevated to 796.8 pg/mL (normal range: 0–97.3 pg/mL). The plain chest radiography showed mild cardiomegaly and no active lung lesion. Transthoracic echocardiography revealed a tumor in the right atrium that appeared lobulated with hypoechogenic internal areas. The mass was initially misdiagnosed as atrial myxoma. Echocardiography also revealed a large, mobile mass extending from the IVC into the right atrium and ventricle, without evidence of attachment to the right atrial or ventricular walls (Fig. 1A). A cardiac CT scan confirmed the presence of an ovoid mass in the right chambers of the heart and in the inferior vena cava, with extensive thrombosis extending to the confluence of the common iliac veins, and with fur-

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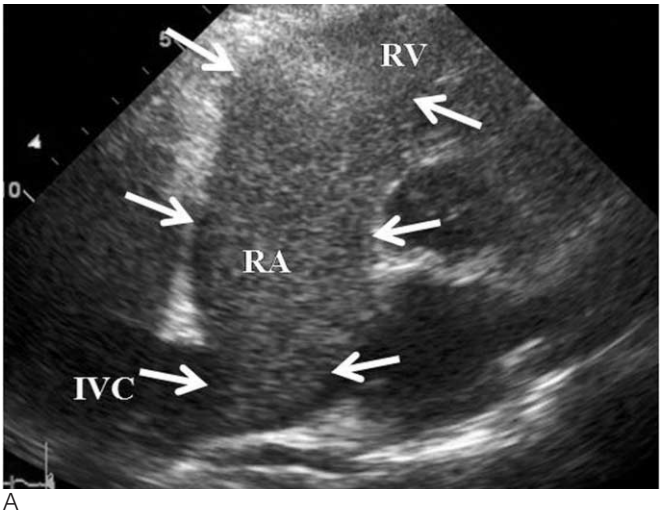

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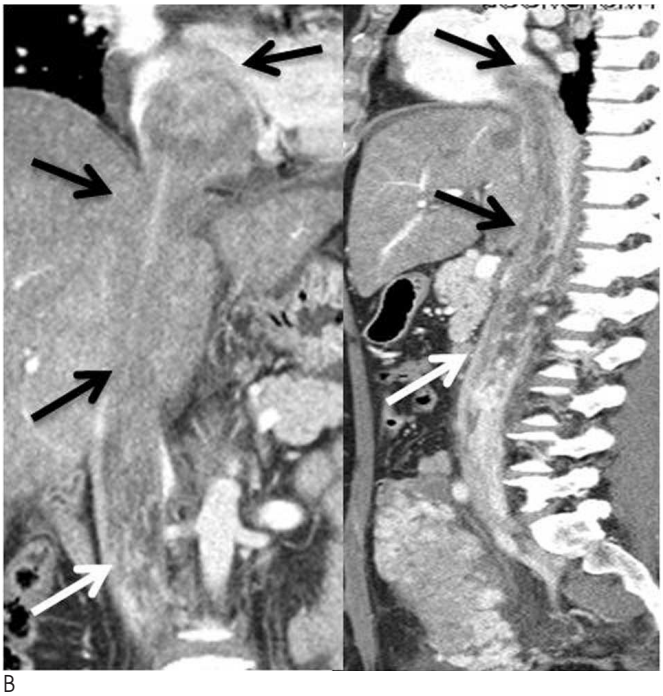

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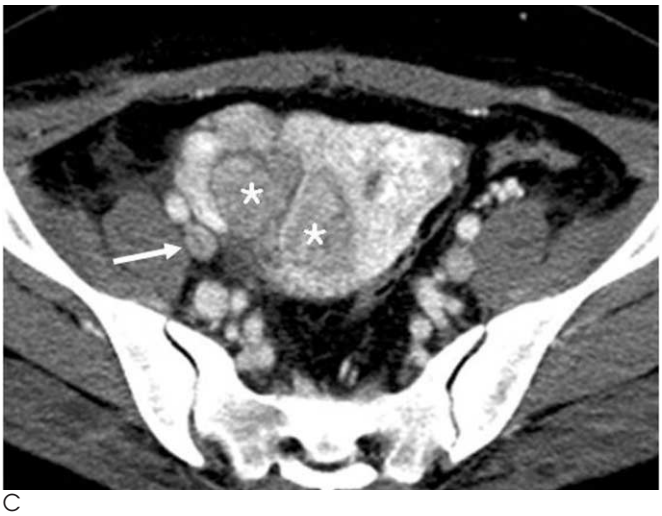

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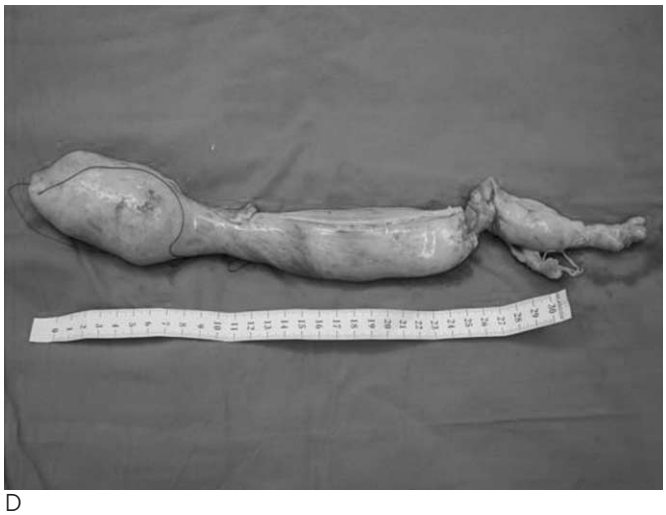

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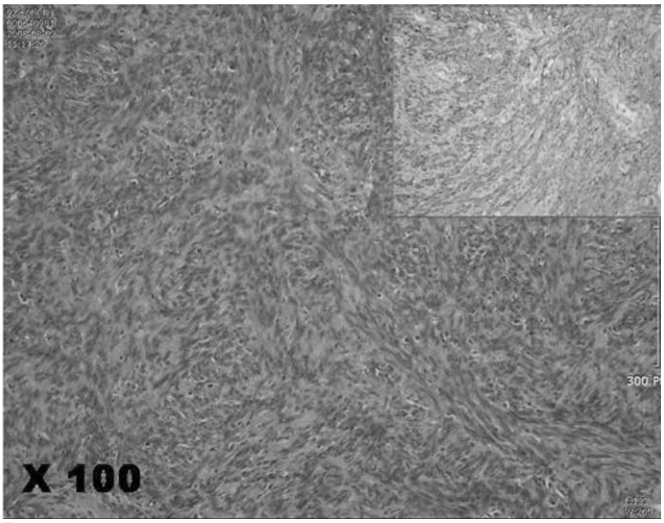

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Fig. 1. A 35-year-old woman with leiomyomatosis involving the IVC and heart.

A. Transthoracic echocardiography demonstrated a large homogeneous echogenic mass (arrows) extending from the IVC into the right atrium and ventricle and it fully occupied these heart chambers.

B. The coronal (right) and sagittal (left) multiplanar reformation images showed a heterogeneous enhancing mass (arrows) involving the right atrium and IVC.

C. The axial pelvic CT scan demonstrated multiple encapsulated masses (asterisks) in the uterus and a dot-like filling defect, which was suspected of being thrombus, in the right external iliac vein (arrow).

D. The tumor specimen from the IVC showed a well circumscribed grey-white mass with a firm, rubbery consistency and it measured 22.5 × 4.5 × 2.5 cm.

E. H & E staining revealed elongated spindle cells arranged in interlacing fascicles (H-E, × 100). The immunohistochemistry revealed strong cytoplasmic staining for desmin (insert image, × 200).

ther thrombotic material inside the right uterine vein (Fig. 1B). In addition, heterogeneously attenuated solid masses and multiple cystic lesions were also found in the uterus (Fig. 1C). Pelvic MRI revealed uterine leiomyomas and the presence of complex solid and cystic masses (not shown).

Surgery was performed over two sessions in the same day. Median sternotomy was performed. The right atrium and inferior vena cava were opened and a reddish-beige smooth, ovoid mass was removed from the right cardiac chambers and the IVC (Fig. 1D). In addition, through another incision that was made at the right lower abdominal quadrant, tumor thrombi were extracted from within the IVC and the right common iliac vein. Seven days after median sternotomy, the patient underwent a follow-up CT scan, which revealed the absence of any residual abnormal mass inside the cardiac chambers (not shown). The postoperative period was uneventful, and the patient was discharged from the hospital on the 10th day after undergoing median sternotomy.

The histologic specimens from the right heart and IVC were diagnosed as leiomyomatosis without any features of malignancy. Grossly, the IVC mass was a well-circumscribed, grey-white tumor with a firm, rubbery consistency, it weighed 163 gm and it was 22.5 × 4.5 × 2.5 cm in size (Fig. 1D). The cut surface showed a whorled appearance with cystic changes. Histologically, the tumor consisted of intersecting fascicles of spindle cells with eosinophilic cytoplasm and bland, elongated nuclei with blunted ends (Fig. 1E). Mitotic features were rare. The immunohistochemical staining showed that the tumor cells were positive for desmin (Fig. 1E, inlet) and actin, but they were negative for CD34 and S-100 protein. All the resected margins were tumor free. Hysterectomy was not performed as the surgeon chose a two-stage operation because of the invasiveness of the combined cardio vascular/gynecological approach and the risk of bleeding due to the systemic heparinization. At three months post-surgery, the patient has been symptom free and doing well without any evidence of tumor recurrence.

Discussion

IVC leiomyomatosis is rare tumor that predominantly occurs in women (1-3), and almost all these patients have a history of hysterectomy or myomectomy, like our patient. The symptoms of these patients occur secondary to the vascular extension and obstruction by the tumor. Two mechanisms have been proposed to explain the spread and growth of tumor into large vessels. The first concerns the direct invasion into the intima of the venous sinuses of the myometrium by the leiomyomatous cells that originate from the uterine myometrium, and the second concerns the proliferation of smooth muscle cells in the venous walls of the uterine veins or pelvic veins (4, 5). In rare cases, the intravenous leiomyomas extend to the right cardiac chambers and this can cause various symptoms that range from dyspnea (as in our case), tachycardia, sudden death, cardiac tamponade and arrhythmias to abnormal bleeding, edema of the inferior limbs and even to Budd-Chiari syndrome by the tumor increasing the blood pressure (6). Therefore, an early and accurate diagnosis is crucial for a good prognosis. IVC leiomyomatosis is usually diagnosed by echocardiography, CT and MRI (7), and the operative strategy can be determined by cardiac catheterization and venography of the IVC. In addition, transabdominal or transvaginal ultrasonography and pelvic MRI can be used to locate the primary site of the tumor. In our described case, the cardiac tumor was initially detected by echocardiography and the intravenous uterine leiomyomatosis with the IVC and intracardiac extensions was confirmed by cardiac CT, whereas pelvic MRI localized the uterine leiomyoma and demonstrated its benignity. Surgery is the treatment of choice for leiomyomatosis and can be performed using a single or double-stage procedure. Complete resection of the tumor and thrombus should be achieved due to its propensity to recur despite its benign nature (8-10). The described case is a typical, but interesting example of intravenous uterine leiomyomatosis with IVC and intracardiac extensions, and we diagnosed all this according to the echocardiographic, cardiac CT and pelvic MRI findings.

References

1. Armstrong PJ, Franklin DP. Pararenal vena cava leiomyosarcoma versus leiomyomatosis: difficult diagnosis. *J Vasc Surg* 2002;36:1256-1259
2. Kasar-Seibert DJ, Gauvin GP, Rogoff PA, Vittimberga FJ, Margolis S, Hilgenberg AD, et al. Intracardiac extension of intravenous leiomyomatosis. *Radiology* 1988;168:409-410
3. Kutay V, Tuncer M, Harman M, Ekim H, Yakut C. Intracardiac extension of intravenous leiomyoma. *Tex Heart Inst J* 2005;32:232-234
4. Merchant S, Malpica A, Deavers MT, Czapar C, Gershenson D, Silva EG. Vessels within vessels in the myometrium. *Am J Surg Pathol* 2002;26:232-236
5. Nam MS, Jeon MJ, Kim YT, Kim JW, Park KH, Hong YS. Pelvic

leiomyomatosis with intracaval and intracardiac extension: a case report and review of the literature. *Gynecol Oncol* 2003;89:175-180
6. Arinami Y, Kodama S, Kase H, Tanaka K, Okazaki H, Maruyama Y. Successful one-stage complete removal of an entire intravenous leiomyomatosis in the heart, vena cava and uterus. *Gynecol Oncol* 1997;64:547-550
7. Kaufman LB, Yeh BM, Breiman RS, Joe BN, Qayyum A, Coakley FV. Inferior vena cava filling defects on CT and MRI. *AJR Am J Roentgenol* 2005;185:717-726
8. Ma L, Wu S, Zou Y, Li W, Gong L, Gerelle G, et al. One-stage surgery of low-grade malignant intravenous uterine leiomyosarcomatosis with right atrium extension. *Ann Thorac Surg* 2007;84:644-647
9. Gaudino M, Spatuzza P, Snider F, Luciani N, Cina G, Possati G. Surgical management of a uterine leiomyoma extending through the inferior vena cava into the right heart. *Heart Vessels* 2002;17:80-82
10. Castelli P, Caronno R, Piffaretti G, Tozzi M. Intravenous uterine leiomyomatosis with right heart extension: successful two-stage surgical removal. *Ann Vasc Surg* 2006;20:405-407

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하대정맥과 심장 내로 퍼진 정맥 내 자궁평활근종증[1]

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자궁에서 기시해서 우심실, 우심방, 하대정맥과 장골정맥을 이환하는 정맥 내 평활근종증은 드문 질환이다. 저자들은 35세 여성에서 심장과 하대정맥 내 종양으로 나타난 평활근종증을 보고하고자 한다. 경흉부 심장초음파, 심장 CT와 골반 내 자기공명영상 소견으로 진단이 이루어졌다.