A rare case of intravascular leiomyomatosis from the ovarian vein to the right atrium in an asymptomatic woman

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Intravascular leiomyomatosis (IVL) is a rare benign condition characterized by non-tissue-invasive intravascular proliferation of smooth muscle cells originating from uterine venous wall or uterine leiomyoma, affecting premenopausal women, typically with a history of uterine leiomyoma or gynecological surgery [1]. The vascular spread is usually *via* iliac or ovarian veins [2], occasionally extending into the inferior vena cava and right heart chambers (intracardial leiomyomatosis).

Symptoms, if present, are usually non-specific and arise from vascular or intracardiac obstruction, potentially leading to cardiac failure. The treatment of choice is total surgical resection, including hysterectomy [3]. However, the therapeutic plan depends on the patient's clinical status, her desire to preserve fertility, and the size and extent of the lesion. Alternative treatment options are hormonotherapy or observation. The recurrence rate is about 16.6%–30% [4], therefore, long-term follow-up is recommended.

This condition was first described by Birch-Hirschfeld in 1896. The incidence of this disease is 0.25% to 0.40% of patients with uterine fibroma [3]. Full IVL pathogenesis remains unclear. Until now, more than 300 cases have been described in the literature.

In this report, we present a case of an asymptomatic 64-year-old woman who was referred to our hospital for further investigation of a mass in the right heart atrium and the inferior vena cava on echocardiography. She had no relevant medical history, and other examinations were negative, apart from the intravascular mass. These included gynecological examination with ultrasound, with no fibroids found. There was no history of gynecological surgery. The differential diagnosis of the mass included thrombus, leiomyosarcoma, soft-tissue sarcoma, lymphoma, tumor thrombosis, and metastasis.

Contrast-enhanced computed tomography (CT) demonstrated intravascular tortuous enhancing non-invasive mass extending from the right ovarian vein through the inferior vena cava to the right atrium (Figure 1A). 18F-FDG positron emission tomography/CT demonstrated low glucose metabolism of the intravascular mass (Figure 1B–C). Both examinations made IVL the most likely diagnosis, the sole confounder was the absence of fibroids or gynecologic surgery. Examinations also ruled out other complications such as organ ischemia.

The multidisciplinary team suggested surgical treatment. The patient consented and underwent a one-stage extensive surgery performed by an experienced cardiovascular surgery team. We administered extracorporeal circulation, deep hypothermia 28°, aortic cross-clamp (for 28 minutes, cardioplegia custodiol 1000 ml), and 8-minute circulatory arrest. During that time, the tumor was extracted in one piece through a radial incision in the right atrium. The inferior vena cava was inspected, and the right ovarian vein was extirpated (Figure 1D). With a partial clamp on the right atrium, we started cardiopulmonary bypass and rewarming, and we sutured the right atrium. No periprocedural complications were reported, and the patient made an excellent postoperative recovery.

Histopathology showed (Figure 1E–F) epitheloid cells with abundant stromal hyalinization immunoreactive for both α-actin and estrogen receptors, consistent with the

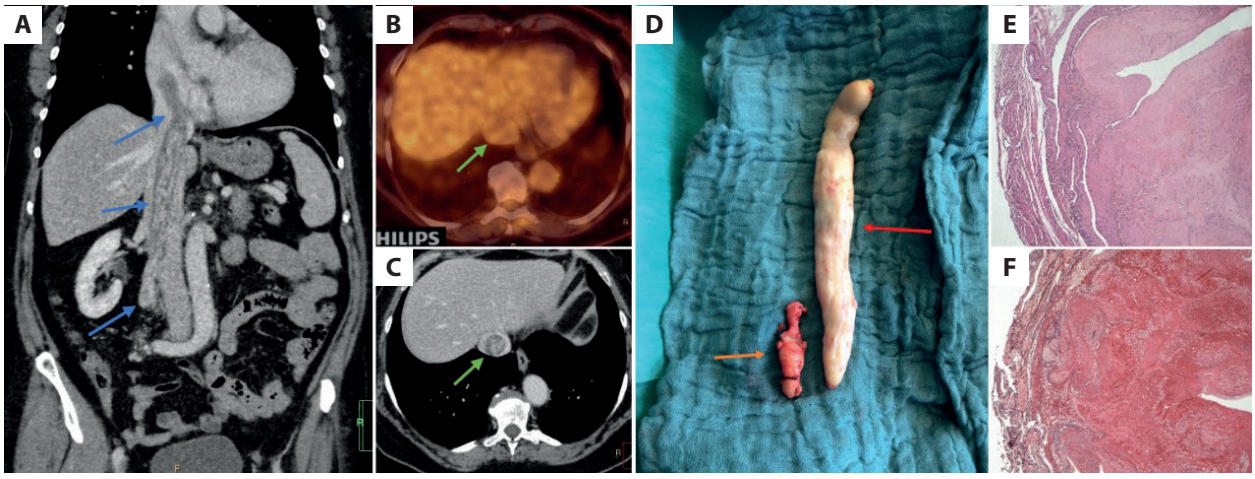

Figure 1. A. Computed tomography (CT) oblique coronal reformat demonstrating enhancing mass extending from the right ovarian vein, through the inferior vena cava to the right atrium (blue arrows). **B, C.** Comparison of an axial slice of contrast-enhanced CT (**B**) and fused positron emission tomography/CT (**C**) demonstrating low glucose metabolism of the intravascular mass (green arrows). **D.** Surgical specimen of the tumor extracted from the inferior vena cava (red arrow) and the extirpated right ovarian vein with tumor (orange arrow). **E, F.** Histopathologic specimen of leiomyoma consisted of epitheloid cells with abundant stromal hyalinization and edema obturating the lumen of the ovarian vein. Hematoxylin and eosin stain (**E**). Verhoeff-Van Gieson stain (**F**)

diagnosis of intravascular leiomyomatosis. Adjuvant hormonal therapy was not indicated since complete surgical resection was achieved [5].

Although the guidelines suggest bilateral salpingo-oophorectomy and hysterectomy to prevent recurrence, the patient preferred regular follow-up without any additional surgery. Subsequently, an expert gynecological ultrasound discovered one small uterine fibroid (9 mm in size). Follow-up examinations (cardiac magnetic resonance, CT, and pelvic ultrasound) showed no signs of local recurrence, and the patient remains asymptomatic.

Article information

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