

Case Report

Intravenous Leiomyomatosis of the Uterus with Intracardiac Extension

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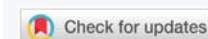
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Abstract

Background: Intravascular Leiomyomatosis (IVL) is an often misdiagnosed rare benign mesenchymal tumor characterized by the presence of vascular extension and invasion of smooth muscle cells in a serpiginous-like pattern, first originating in uterine smooth muscle cells. Its growth pattern can involve both ovarian veins, the inferior vena cava, and even reach the right atrium/ventricle in 45% of the cases. The incidence has been reported to be 0.25 to 0.40% of patients with uterine leiomyoma, with about 300 cases reported in the literature. Also, since the tumor is hormone-dependent, most affected individuals are premenopausal women in middle age. Optimal treatment for IVL is complete surgical removal with hysterectomy and oophorectomy, independent of stage. The most frequent perioperative complications are hemorrhage due to tumoral hypervascularization, embolism, and the usual laparotomy complications. We present the case of a 51-year-old female with IVL stage 3 with complete single-stage surgical resolution.

Introduction

Intravascular Leiomyomatosis (IVL) is an unusual type of leiomyoma beyond the uterus; it originates in uterine smooth muscle cells, and some of the other extrauterine forms of benign metastasizing leiomyomas include diffuse peritoneal leiomyomatosis, retroperitoneal, and parasitic leiomyomas [1-9]. Since the tumor is hormone-dependent, most affected individuals are pre- or perimenopausal women, ages 40-50. History of hysterectomy or uterine leiomyoma may be present [10,11]. The incidence of IVL is 0.25% - 0.40%. Growth into the veins of the myometrium and parametrium occurs in 80% of the cases, and extension to the heart in 10% - 45% [11-13].

According to the Mendelian randomization study by Qu, et al. [2], nulliparity, hypertension, obesity, high insulin/

glucose levels, and endometriosis are risk factors, while multiparity, late menarche, and polycystic ovary syndrome may have a protective role against uterine leiomyomas [2].

They are influenced by both dietary factors and genetic mutations. A diet high in red meat and low in fruits, vegetables, and dairy products is associated with an increased risk of developing fibroids. Additionally, vitamin D deficiency has been linked to larger and more numerous fibroids. Conversely, a healthy, balanced diet may offer a protective effect by modulating estrogen activity and reducing inflammation.

On the genetic level, mutations in the MED12 gene are the most frequently observed, present in about 70% of the patients with this pathology. These mutations affect gene transcription and promote abnormal cell growth. Other significant genetic

alterations include chromosomal rearrangements involving regions such as 12q14 15, 6p21, 7q22, and 1p36, changes in the HMGA2 gene, and mutations in the Fumarate Hydratase (FH) gene are found in hereditary leiomyomatosis [3].

The most common symptoms and signs are dysmenorrhea, palpable pelvic masses, ascites, right heart failure, syncope, and even sudden death [4,5,14]. Different stages of the disease have been described as follows: stage 1) confined to the pelvic cavity, stage 2) extension to the iliac vein or the inferior vena cava below the renal vein, stage 3) extension to the vena cava above the renal vein, including the right cardiac cavities, 4) tumor invading the pulmonary arteries. Computed tomography (CT) and Magnetic resonance (MRI) are the principal imaging modalities for the initial evaluation of IVL, CT commonly demonstrate low-attenuation intraluminal filling defects within venous structures frequently associated with this pathology, however MRI in particular provides superior soft tissue contrast and multiplanar imaging capability, which are essential for accurately see the extent of the tumor involvement [12,14].

Surgical approach consisting in a complete surgical removal with hysterectomy and oophorectomy independent of stage is the optimal choice of treatment; it's still in debate whether it is better to have a single-staged versus two-staged surgery, but a few studies recommend one-staged approach to avoid the risk of tumor or thrombus embolization and tumoral, this one could performed to remove the entire tumor, provided there is no invasion of the cardiac chambers, because when the right atrium is involved the amount of surgical time required increases as well as the risk of complications. However, no specific approach has been demonstrated to be superior [6].

Case report

A 51-year-old woman with the following relevant medical history: previous cardiac surgery due to auricular neoplasm one and a half years before the actual event, with a histopathologic report of myxoma. At the time of evaluation, the patient reported abdominal pain, nausea, and sensation of sudden death on two occasions; bilateral leg oedema was observed. An angiotomography was performed with the finding of a hypervascularized pelvic tumor with drastic captation of contrast in the arterial phase, beginning in the uterine fundus, and making its way to the right gonadal vein towards into the inferior vena cava and going upward into the right atrium (Figure 1). Blood tests only highlight the abnormal elevation of CA125.

Going further with the evaluation, a multidisciplinary approach was made, including vascular surgery, cardiovascular surgery, gynecology, and interventional radiology. As a result, all of them concluded that the angiotomography and clinical findings were compatible with IVL, ruling out the possibility of sarcoma. During this clinical workup, a transesophageal echocardiogram was solicited and performed, with the

findings of right auricular enlargement with a mobile intracavitary mass causing tricuspid partial obstruction due to direct contact with the valve leaflets, which impose a risk of sudden death.

With the evidence obtained, it was decided to admit the patient to the coronary care unit and to plan a multidisciplinary strategy, which was developed as follows:

The first step was endovascular embolization of both uterine arteries and the nutritional tumor artery with a mixture of lipiodol and hystoacril 10:1 and three coated 6 x 10mm and 6 x 20mm coils in each uterine artery; control angiography showed successful tumor embolization and uterine artery occlusion (Figure 2).

The second step was the one-stage surgical intervention, beginning at the same time as a laparotomy for hysterectomy and bilateral oophorectomy, and a sternotomy to establish

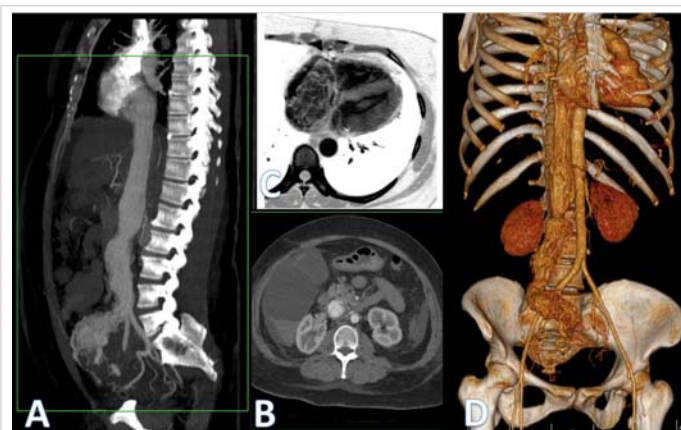


Figure 1: A) Sagittal CT reconstruction showing vascular extension of the uterine leiomyoma into the inferior vena cava and right atrium. B) Axial CT showing an enhanced arterIALIZED mass within the inferior vena cava. C) Negative CT showing a saccular morphology of the tumor inside the right atrium. D) 3D reconstruction demonstrating the right uterine artery as the main nutritional vessel of the tumor.

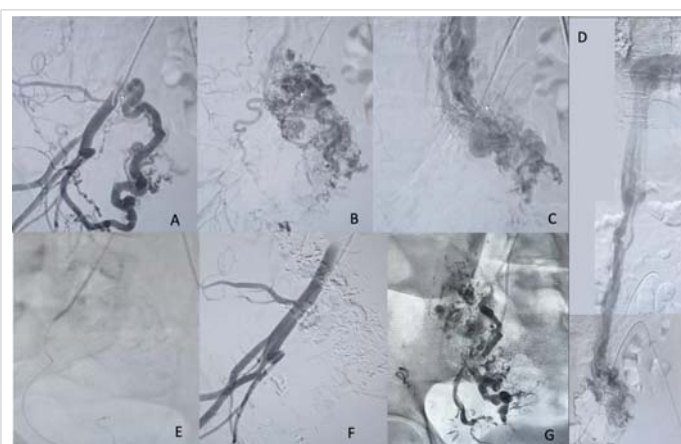


Figure 2: A) Digital subtraction angiography of the right uterine artery showing a hypervascular tortuous tumor (B) with infiltration of the right ovarian vein (C) and its extension to the inferior vena cava and right auricle (F). (D) Selective cannulation of the ascending portion of the right uterine artery. E) Digital subtraction angiography showing complete occlusion of the tumor and the right uterine artery. F) Single-take radiograph showing hyperdense fluid emboli within the tumor.

cardiopulmonary bypass for circulatory support (Figure 3). When hysterectomy and bilateral oophorectomy was completed, the ovarian vein containing the tumor was dissected until its junction to the inferior vena cava, proximal and distal vascular control was obtained, an incision in the ovarian vein was made and with gentle traction the tumor extraction was completed (Figure 4). Once tumor extraction and hemostasis verification were assured, circulatory support was removed, abdominal Blake drains, and retrosternal chest tubes were placed prior to closure.

This surgical intervention lasted 4 hours, with a cardiopulmonary bypass of 108 minutes, without aortic clamping and blood loss of 4000 ml. The histopathologic report was: leiomyoma with myxoma component (Figure 4).

Discussion

Intravascular Leiomyomatosis (IVL) is a rare benign tumor originating from a uterine leiomyoma or the venous wall of uterine veins. Although it is histologically benign, its behavior is aggressive, resembling that of a malignant tumor. IVL can be localized into the uterus, but in some cases, it may extend to venous structures close to the uterus such as the gonadal vein or iliac vein, and in rare cases, it uses these vascular structures to reach the vena cava, where it may extend all the way into

the right atrium and even the pulmonary artery. Despite its extensive growth, the tumor appears to be limited to the intravascular/intracavitary space, with no actual invasion of the vascular wall [4].

The exact physiopathology of IVL is unknown; however, there are two prominent theories: the first one suggests that leiomyoma originates from the uterus and later grows into the vasculature, and the second, that it originates directly from the vessel wall. In this case, the tumor originated from the uterus, traveled inside the uterine vein to the inferior vena cava, and then to the heart. Imaging evaluation of this pathology is initially done by CT, where it has been described as filling defects within the venous structure or as a hyperattenuating intravenous mass. It can also demonstrate the direct continuity between the intravascular tumor and the pelvic venous system. The 3D reconstruction is useful to show the serpiginous-like growth pattern in cases where the tumor reaches any of the right cardiac chambers. On MRI, the intravascular component is characterized by a mass of dilated, convoluted tubular structures with a solid configuration, often described as a worm-like mass. Signal characteristics on T1-weighted images are generally isointense to mildly hyperintense relative to skeletal muscle, whereas T2-weighted sequences typically show moderate to marked hyperintensity. These imaging features reflect the tumor's histopathological composition, particularly the relative abundance of smooth muscle cells and fibrotic, hyalinized vascular elements. [12,14]. This pathology may have some imaging differential diagnoses like bland thrombus, but in this context, the intraluminal material does not enhance, and it can be easily excluded in contrast media phases on CT or MRI, even though this one only occurs in some specific cases like post-operative state or chronic debility, all of which are absent in our case [12]. Another one is malignancy like leiomyosarcoma arising from the wall of the IVC on the basis of imaging features alone, but the main difference could be the growth patterns associated to this tumor: extraluminal (62%) intraluminal (5%) and mixed type extra and intraluminal (33%) involvement, however, histopathologic analysis is necessary for the diagnosis confirmation. In the context of a cardiac mass, myxoma should be considered a differential diagnosis, but given that the right atrium is not a common location for a myxoma (10% of the cases), the presence of a right atrium mass with caval involvement in a woman could increase the possibility of IVL [7,8].

Although histologically it is a benign tumor, it can be considered aggressive, since it has high rates of recurrence and can have fatal consequences due to its ability to vascular blood vessels.

Macroscopically, the cut surface is whitish-gray or pinkish-white and molded on withdrawal from the affected vessel, which usually originates from the uterus and may extend to the heart.



Figure 3: A. Endovascular leiomyomatosis. B. Uterus with polypoid mioma. C. Distal uterine artery with endovascular tumor.

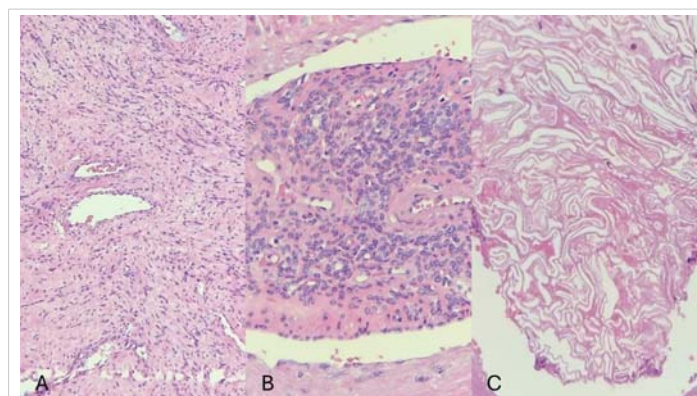


Figure 4: A. Hematoxylin-eosin 10x. Uterine leiomyoma with spindle cells without atypia or mitosis. B. 40x with intravascular spindle cells in vessels of the tubal hilum. C. Presence of liquid embolic material (lipiodol/hystoacril) within the tumor vessel.

Histologically, it is characterized by smooth muscle proliferation within the vessels or lymphatic vessels with little detectable cellular changes or increased cellularity, epithelioid morphology, myxoid and mucoid changes, sometimes papillary appearance, and few mitoses [9].

By immunohistochemistry, the positive markers Vimentin, smooth muscle actin, estrogen and progesterone receptors, CD10 positive focal or negative, and CD34 expression indicate smooth muscle origin, and Ki67 is less than 5%. Differential diagnoses include low-grade endometrial stromal sarcoma, uterine leiomyosarcoma with vascular invasion, and cotyledonian dissecting leiomyoma (Stenberg tumor) [9].

Mutations have been found on chromosomes 1p, 22q.2q and 14q as well as 10q22.2, 10q24.32, 13q14, and 13q21-31 that will help to more correctly identify the lesions and differentiate them from other histologically similar lesions [15].

Surgical classification of IVL is based on the anatomical extension of the tumor, stage 1: confined to the pelvic cavity, stage 2: extension to the iliac vein or the inferior vena cava below the renal vein, stage 3: extension to the inferior vena cava beyond the renal vein, including the right cardiac cavities and stage 4: tumor invading the pulmonary arteries.

The common presenting symptoms are related to uterine disease: abnormal uterine bleeding, pressure, and lower abdominal pain. However, if invasion of the inferior vena cava is extensive, it can also present as lower extremity venous insufficiency and abdominal pain. Due to the tumor extension over the IVC, it can cause some complications as Budd-Chiari syndrome or other secondary pathologies, such as tricuspid valve stenosis that will result in tricuspid insufficiency, heart failure, and even sudden death due to incarceration of the tumor in the valve [5].

Surgical treatment appears to be the only treatment with a curative purpose. Differences in the extent of surgical resection vary; it must always include hysterectomy with or without oophorectomy is an essential part of treatment. Oophorectomy is recommended since some studies have shown a higher risk of recurrence when it's omitted [6].

More extensive surgical intervention may be divided into one-step or two-step approaches. A one-step approach involves a thoracotomy and laparotomy in a single surgical event, with hysterectomy and resection of the intravenous and intracardiac portion of the tumor. This surgical approach allows for optimal tumor resection, and if needed, can bring appropriate exposure for vascular reconstruction and cardiac exploration. Some other points in favor of a single-step procedure are avoidance of a second anesthesia induction, less risk of progression or tumor embolization, and lower costs. However, the extended anesthesia time and surgical trauma are downsides to this kind of approach and may not be ideal for every patient.

The two-step approach can be divided into thoracotomy first or laparotomy first. The majority of these interventions begin with resection of intracardiac leiomyoma, with cardiopulmonary bypass support. In a second surgical procedure, the hysterectomy and oophorectomy remove pelvic leiomyoma. Resection of the vascular portion of the leiomyoma can be done in either intervention [6].

Presurgical procedures to reduce the tumor and obstruct the blood flow have been proposed, including uterine artery embolization and premedication with goserelin. We considered the use of goserelin, but knowing the risk of sudden death, we favored the embolization option.

Not much is found in the literature about endovascular approaches for tumor resection, in one of those cases it is mentioned a percutaneous extraction of a tumor is mentioned as if it were a venous thrombus. There are no reports where an endovascular approach to embolize the tumor is combined with open surgical resection.

In cases of incomplete resection, medical therapy has been used to reduce recurrence. This is based on the theory that IVL has similar histological characteristics to uterine leiomyomas. The objective of medical therapy is to reduce estrogen levels, thus reducing the growth stimulus for the tumor. Various studies have followed the effect of postsurgical medical treatment, including selective estrogen receptor modulators (SERM), progesterone, which did not have a significant impact on recurrence; however, it appears that gonadotropin-releasing hormone agonists prevent recurrence when given for 6 months postoperatively.

Incomplete resection has a recurrence rate of 30%; factors that influence the likelihood of recurrence include the resection of the ovaries.

In this case, we have done the complete resection in a one-stage surgery with previous endovascular embolization.

Ethical consideration

This clinical case was prepared in accordance with the ethical principles established in the Declaration of Helsinki. Written informed consent was obtained from the patient for the collection, analysis, and publication of relevant clinical information and images, ensuring the confidentiality of her personal data at all times. The report was reviewed and approved by the corresponding institutional ethics committee, considering that this is a minimal-risk study as it did not intervene in the course of treatment nor modify medical decision-making. Care was taken to ensure that the presentation of the case does not allow patient identification.

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