

UTERINE LEIOMYOMATOSIS WITH INTRACARDIAC EXTENSION: ONE-STAGE SURGICAL APPROACH

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Abstract

Intravenous leiomyomatosis is a rare benign vascular tumor. Intracardiac extension is infrequently but can lead into a threatening-life situation.

We report a 41-year-old woman who has undergone previous hysterectomy due to uterine myomatosis and now presents with a pelvic mass contacting the venous system through the right internal iliac vein and extending up to the right pulmonary artery. Surgical resection of the pelvic mass and intravenous tumor removal was successfully performed in a single-stage approach with cardiopulmonary bypass. Patient recovered satisfactorily, being asymptomatic at hospital discharge time.

One-stage surgical approach needs a multidisciplinary surgical team and needs a longer operative time than two-stage approach. However it can be a safety treatment option for patients with good general condition. Furthermore cardiopulmonary bypass guarantees a safe procedure, avoiding renal and hepatic ischemia and potential embolizations into pulmonary circulation during mass or ilio cava venous sector manipulation.

Keywords: *Leiomyomatosis; Uterine Neoplasm; Cardiac Imaging Techniques; Vascular Surgical Procedures; Venous Thrombosis; Cardiopulmonary Bypass.*

INTRODUCTION

Intravenous leiomyomatosis (IVL) is a rare benign vascular tumor that was first described in 1897 by Birsch-Hirschfeld¹. However it was not until 1907 when the first intracardiac extension was reported by Durk and Hormann². Up to 45% of cases this neoplasm extends to iliac veins or inferior cava vein (IVC) but intracardiac extension is less frequent with only 6 to 30% of the reported cases³.

CLINICAL CASE

A 41-year-old nulliparous woman presents with costal pain and hemoptysis after a bicycle trauma. The patient had undergone hysterectomy 5 years ago due to uterine myomatosis.

Computed Tomography Angiography (CTA) shows a right anexal mass of 6x5 cm with thrombus extending from the right pulmonary artery into the IVC and even reaching right common iliac vein (Figure 1). A thoracic CTA showed a pulmonary thromboembolism affecting right pulmonary

artery, a complete occlusion of the inferior lobar artery and partially occlusion of the mid lobar artery. Because of this, anticoagulation was started at the moment of the admission with low-molecular-weight heparins.

In order to determine the precise extent of the thrombus, we perform a transesophageal echocardiogram where we noted thrombus extending from superior and inferior vena cava into the right atrium and reaching the pulmonary artery trunk. Bronchoscopy showed no pulmonary involvement.

Magnetic Resonance Imaging (MRI) confirmed the previous CTA findings, with the intravenous mass at right pulmonary artery, superior and inferior vena cava, right common iliac vein and also contacting the right anaxal mass through the right internal iliac vein (Figure 2). This intrapelvic tumor presented several engorged veins with total lumen obliteration.

An echocardiogram was performed and excluded any kind of alteration of the pulmonary and tricuspid valves.

After addressing the case in multidisciplinary team, including professionals from Radiology, Cardiac Surgery, Gynecology and Vascular Surgery departments, we decided to perform an anaxal mass resection with intravenous thrombus removal during the same procedure. In this way the surgical procedure was carried out by gynecologists, cardiac and vascular surgeons in conjunction.

We started with a simultaneous sternotomy and xipho-pubic laparotomy. First of all the pelvic mass was removed, with resection of both ovaries and the right Fallopian tube (Figure 3). Tissue dissection and mobilization was very laborious due to fibrosis caused by prior hysterectomy and also because the presence of several engorged veins. The tumor had a soft consistency. In order to obtain a complete resection, the right internal iliac vein and the right gonadal vein were both sectioned to avoid tumoral progression.

Subsequently, the patient was started on cardiopulmonary bypass (CPB) which involves cannulation of ascending aorta and superior vena cava (Figure 4). The inferior vena cava cannula was placed just proximal to the right atrium. With vascular and cardiac surgeons working simultaneously a right atriotomy, lower cavotomy and venotomy on the right common iliac vein were performed (Figure 4). Tumoral thrombus in the IVC and right common iliac vein was easily removed but the right atrial thrombus was more adherent and could not be removed by simple traction. In this moment we proceed to open the right pulmonary artery to ease execution. The extracted thrombus was filiform and with small dilations, just like rosary beads (Figure 3). All vascular sutures were performed with direct suture using polypropylene of differing thickness according to surgeons preference. 23 minutes of cardiac arrest at 25°C were needed for the procedure. The profound hypothermia and low CPB output was deemed

necessary due to the extensive venous recanalization which could present a very serious intra-operator hemorrhage if usual normothermia and CPB output were maintained. Total duration times of intervention and CPB were 230 minutes and 94 minutes respectively.

There were no incidences in the postoperative period. During and after the intervention, a total of 4 packed red blood cell transfusion was performed. A postoperative echocardiogram showed no pulmonary or tricuspid valve alterations.

Treatment with low-molecular-weight heparin continued in the postoperative period and maintained after discharge for 3 months (the patient preferred low-molecular-weight heparin over oral anticoagulation).

The patient stayed at ICU for 24 hours, and hospitalized for 8 days before discharge. During admission there was no need for aminergic support and after 3 years there is no evidence of tumor recurrence.

DISCUSSION

Although IVL is initially considered a benign vascular tumor, when tumor extends up to the right atrium or pulmonary artery it becomes a life-threatening situation⁴. For this reason we must have a high index of suspicion, especially when we face-up with a women who has suffered an uterine leiomyoma (64% of women affected have undergone previous hysterectomies for this reason⁵) and who presents with obstructive venous symptoms or with a mass located at the right atrium. This neoplasm is usually asymptomatic and it's only when an important venous obstruction is established when it normally causes symptoms and is diagnosed^{4, 5}. Main symptoms are those related to venous stasis like low abdominal pain, lower extremity edema or those related to the cardiac system such as dyspnea, chest pain, congestive failure or even sudden death⁶.

There are two theories about etiology of IVL, however none of them are widely accepted. The first one maintains that it's caused as a result of endovascular smooth muscle cell proliferation⁷. The other theory affirms IVL as a result of a previous uterine leiomyoma proliferation⁸.

Regarding histology the main feature is the presence of benign-looking smooth muscle cells into uterine and extrauterine venous system⁷.

The key for an optimal treatment plan its to define accurately the extension of the tumor, reason why imaging tests as CTA or MRI are mandatory. Sometimes it could be difficult to differentiate from other malignant smooth muscle cells in these image tests. Both electrocardiogram and echocardiogram are also important in order to determine mass extension.

Most authors agree that a successful treatment includes complete excision of the mass. In some cases it

can be really difficult to perform a total excision of the tumor, especially when internal iliac veins are affected. Incomplete resections usually result in recurrences⁹. When total excision is not possible adjuvant hormonal therapy with Tamoxifen, Gonadotrophin-releasing hormone (GnRH) antagonists, progestogens or aromatase inhibitors can be helpful in preventing future recurrences¹⁰.

There is some debate about performing surgical intervention in a single-stage or in two-stages, with each approach having its own advantages and disadvantages.

The main advantage of a two-stage approach is a shorter operative time for each surgery and a lower risk of abdominal and pelvic bleeding because systemic heparinization is not required.

In this case we opted for a single-stage approach due to the good general condition that the patient presented, and the extension into the pulmonary artery reinforced the idea that to completely remove the thrombus a right atriotomy was necessary. Moreover, this single-stage approach minimized the risk of catastrophic potential tumor embolization into the distal pulmonar

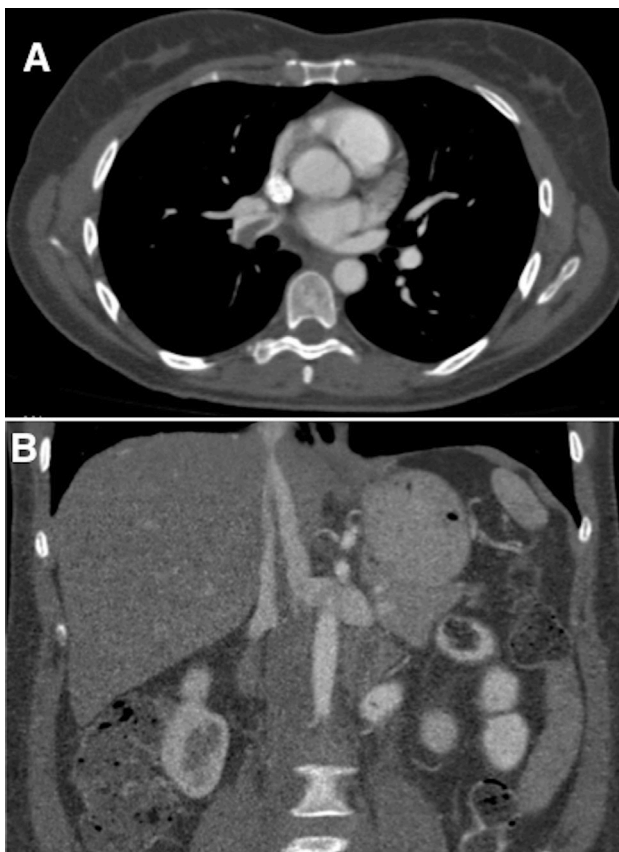


Figure 1

Computed Tomography Angiography (CTA) which shows a right anaxal mass of 6x5 cm and thrombus extending from the right pulmonary artery into the inferior vena cava (IVC) and even reaching right common iliac vein.

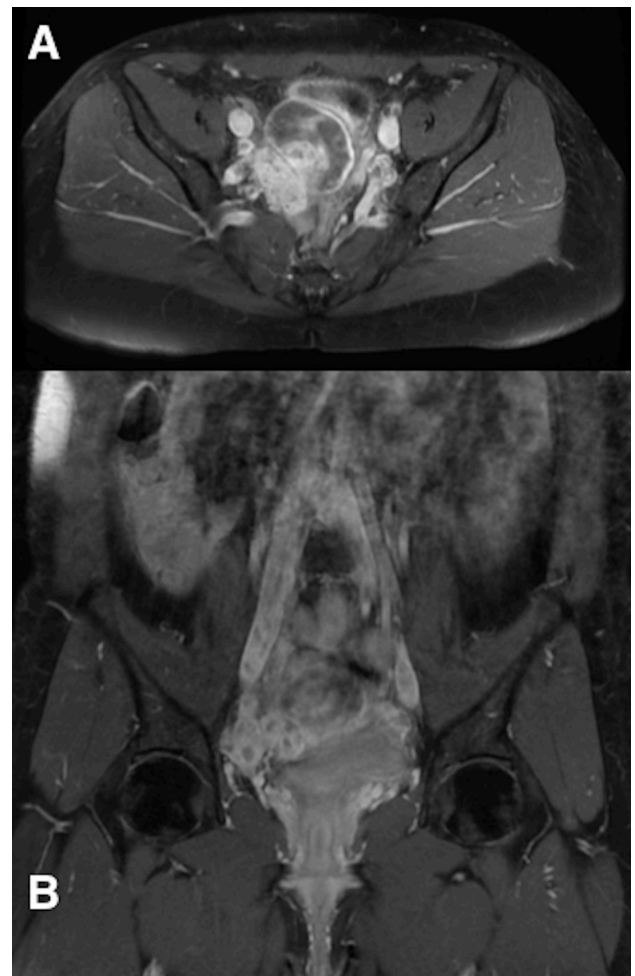


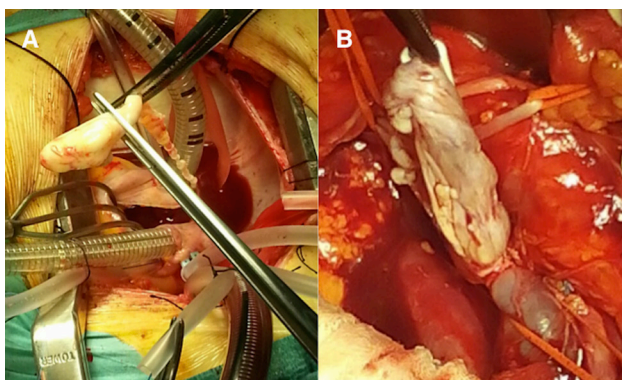
Figure 2

Magnetic Resonance Imaging (MRI) confirming previous CTA findings, with the intravenous mass at right pulmonary artery, superior and inferior vena cava, right common iliac vein and also contacting the right anaxal mass through the right internal iliac vein.



Figure 3

(A) Extracted intravenous thrombus. (B) Pelvic mass which involves both ovaries and the right Fallopian tube.


Figure 4

(A) Thrombus extraction through a right atriotomy with cardiopulmonary bypass (CPB). (B) Venotomy on the right common iliac vein with thrombus inside.

artery bed. Hypothermia was not extreme (25°C) avoiding possible complications derived from its use, but allowing a lower BPC output to diminish the risk of bleeding from the collateralized venous channels. A multidisciplinary approach was essential, first performing careful hemostasis while resecting the pelvic mass with Gynecologists; and thereafter a simultaneous procedure with Cardiac Surgeons allowing shortening operative time that finally lasted 4 hours.

In conclusion we think that CPB guarantees a safe procedure, avoiding potential catastrophic tumor embolization into pulmonary circulation during mass or ilio cava venous sector manipulation and avoiding renal and hepatic ischemia.

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