

# Metastasizing Leiomyoma Obstructing the Right Ventricular Outflow Tract

## Abstract

A very loud systolic murmur was identified during a pre-operative evaluation of a 51-year-old woman for an elective hysterectomy. The TTE showed a 4.7 cm intracardiac mass obstructing the RVOT. The patient was scheduled instead for resection of the mass. Before anesthesia induction, the surgical team and perfusionist were prepared to initiate CPB in case of circulatory collapse. After induction of general anesthesia, the patient became hypotensive, requiring vasopressor support. She recovered and was then successfully placed on CPB. The mass was removed without incident, and a TEE confirmed resolution of the RVOT obstruction. The patient did well post-operatively.

**Keywords:** Benign metastasizing leiomyoma, intracardiac mass, right ventricular outflow tract obstruction

## Introduction

Leiomyomas are the most frequent gynecological tumors. Although the vast majority involve only the uterus, in some infrequent cases, the uterine leiomyoma may have intravenous proliferation, peritoneal dissemination, or distant metastasis.<sup>[1]</sup> The most frequently affected organ is the lung,<sup>[2]</sup> but metastases have been described to paraaortic lymph nodes, abdominal lymph nodes, heart, breasts, and liver among others.<sup>[3]</sup> Although the histology is benign, intracardiac leiomyomas have been associated with sudden death, secondary to obstruction of the ventricular outflow tract.<sup>[4]</sup> We present a challenging case of a benign metastasizing leiomyoma (BML) obstructing the right ventricular outflow tract (RVOT).

## Case History

A 51-year-old pre-menopausal female who presented to emergency department with abdominal pain and abdominal distension was scheduled for resection of a large uterine leiomyoma (19.3 × 14.5 × 16.7 cm). During the pre-operative anesthesia evaluation, she was found to have worsening dyspnea and an intense systolic murmur. Transthoracic echocardiogram (TTE) and a cardiac MRI revealed a 4.7 cm soft tissue mass that was nearly occluding the RVOT and extending into the main pulmonary artery,

a 2.7 cm mass in the pericardium, and a 1 cm nodule within the left ventricular lateral myocardial wall [Figures 1a and b]. Her right ventricular ejection fraction was 41%, the right ventricle (RV) was severely dilated, there was severe tricuspid regurgitation (TR) and the right ventricular systolic pressure (RVSP) was 141 mmHg. Following a multidisciplinary meeting, the patient was scheduled for an urgent resection of the RV mass due to the hemodynamic compromise caused by the RVOT obstruction.

In the operating room (OR), the patient was extremely short of breath, anxious, unable to lie flat, and required left lateral tilt to reduce the aortocaval compression produced by the uterine tumor. Before induction of anesthesia, a 18G femoral arterial line and a 9Fr two-lumen femoral central line introducer were placed. In anticipation of hemodynamic instability, preload was optimized and the patient was positioned in lateral decubitus. Epinephrine, norepinephrine and milrinone infusions were prepared. The cardiac surgeons were gowned, the CPB cannulas were readily available, and the patient was prepped and draped for surgery. Her starting blood pressure (BP) was 170/117 mmHg (MAP: 135 mmHg). After cautious induction with fentanyl, etomidate, succinylcholine and easy intubation, the patient's SBP dropped to 65 mmHg. Boluses of epinephrine were administered, and an infusion

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started. Within 2 minutes, the patient's SBP improved to 100 mmHg. She was cannulated and placed on CPB within 10 minutes of inducing anesthesia.

The mass on the left ventricular wall was approximately 2 cm in diameter, and the left anterior descending coronary artery appeared to be running through it [Figure 2a]. Distal LAD flow was measured in the OR, and there were not EKG or wall motion abnormalities. For this reason, it was decided to biopsy rather than excise this mass. To remove the obstructing RVOT mass, the surgeons accessed the right atrium and retracted the tricuspid valve leaflets. A 4 × 3 cm oval-shaped mass was removed [Figure 2b]. The consistency of the mass appeared to be hard and well formed. A follow up transesophageal echocardiogram (TEE) showed no further obstruction of the RVOT [Figure 3], and the RV dilation, as well as the TR, had improved. The patient was weaned from CPB and the chest closed. She was taken to the critical care unit where she was extubated within the next hour. The patient's post-operative course was uneventful, and she was discharged home on postoperative day eight.

Pathology returned as smooth muscle neoplasm with no mitosis, necrosis nor cellular atypia, positive for desmin, estrogen receptor, progesterone receptors, and a moderate proliferative activity by Ki-67 (10%) which is compatible with a BML.

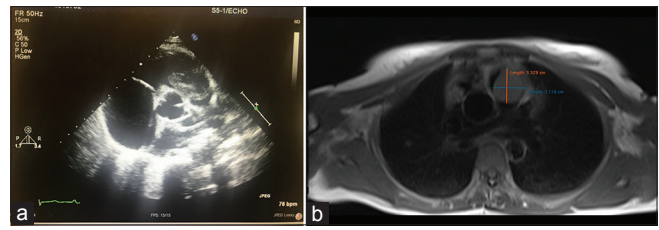
## Discussion

"Benign" is a rare pathology that presents when low-grade smooth muscle neoplasms are present in a distant organ. BML occurs predominantly in women of late childbearing age or older with a mean age of diagnosis of 47.3 years old.<sup>[5]</sup> The determination of BML is based on the immunohistochemical analysis that shows well-differentiated smooth muscle cells with a low proliferation activity. Smooth muscle desmin marker, estrogen receptor, and progesterone receptor are usually positive.

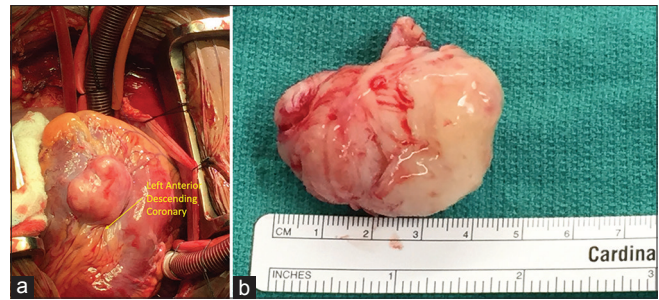
The perioperative management of a BML obstructing the RVOT is exceptionally complex and challenging. If this condition had not been identified before the initially planned hysterectomy, the results could have been catastrophic.

Since the mass was obstructing the RVOT, induction of anesthesia was a critical step. In the scenario of a cardiac arrest, standard resuscitation measures such as chest compressions and infusion of vasoactive drugs would have been ineffective.

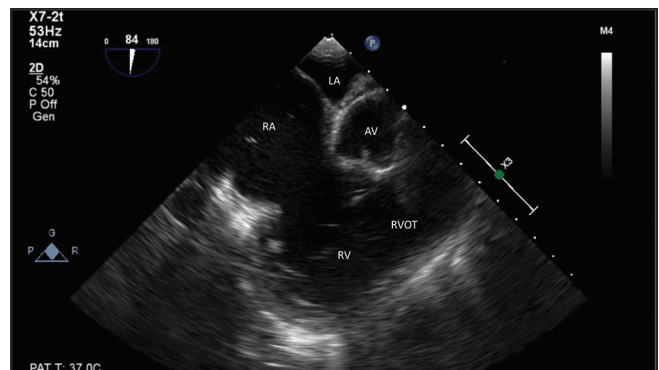
Given the above, the management approach should consider preparing for an imminent circulatory collapse after induction of anesthesia. Adequate adjustment of the preload and minimizing the hypotensive effects of induction agents should be of paramount importance in the preoperative planning of this type of cases. Vasoactive



**Figure 1:** (a) Transthoracic echocardiogram exhibiting the mass located inside the right ventricle. (b) MRI presentation of the leiomyoma in the right ventricle obstructing a significant portion of the RVOT



**Figure 2:** (a) Left ventricular mass with the left coronary artery running through it. (b) 4 × 3 cm mass removed from RVOT



**Figure 3:** Transesophageal echocardiogram Mid-Esophageal RV Inflow-Outflow after intracardiac tumor excision

drugs should be rapidly available, to temporarily maintain the systemic perfusion pressures while awaiting initiation of CPB. A multidisciplinary approach where the surgical team is present throughout induction is essential as rapid implementation of CPB might be needed.<sup>[6]</sup>

Our case demonstrates once again, that a thorough pre-operative evaluation is critical to avoid catastrophes, like the one that could have occurred if this patient had undergone surgery without adequate preparation. A multidisciplinary plan developed jointly with the surgical team in advance is crucial to the success of the procedure.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### **Conflicts of interest**

There are no conflicts of interest.

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