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Case Report

# Intravenous Leiomyomatosis with Intracardiac Extension Using Extracorporeal Membrane Oxygenation in Management Approach

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## Abstract

Little is known about using extracorporeal membrane oxygenation and ECG gated triple rule-out computed tomography to manage and diagnose a large cardiac free wall tumor in a 39-year-old female with Intravenous leiomyomatosis with extension into the right heart.

## Introduction

Intravenous Leiomyoma (IVL) is a rare smooth muscle cell tumor that originates from the walls or uterine venous channels [1]. Although IVL is a benign tumor, the course of the disease can be aggressive. IVL can extend from the uterine vein channels into the right heart chambers via the inferior vena cava vessels, resulting in intracardiac leiomyomatosis (ICL). ICL can lead to complete right ventricle outflow tract obstruction resulting in an increased risk of sudden death [2]. Due to the rarity of this condition, limited literature is available on case reports describing IVL with intracardiac extension. Thus, it is empirical to report cases of IVL with intracardiac extension and discuss clinical presentation, symptom progression, and treatment management. For this case report, we will discuss the use of extracorporeal membrane oxygenation (ECMO) in the management of a case presenting with ICL prior to performing a two-stage surgery removal of the tumor [3].

## Clinical Case

A 39-year-old female presented to primary care physician following a one-week history of ongoing nausea. At the time of presentation, she was discovered to have leukocytosis. An outpatient abdominal computed tomography discovered a large 77 x 40 mm mass in the right atrium. The mass was initially thought to be a large thrombus arising from the inferior vena cava and extending into the pelvis. The patient was transferred to our institution for evaluation of a suspected a cardiac tumor thrombus. Additional history and physical examination in our ED discovered, the patient had also experienced abdominal pain, nausea, mild swelling of the left leg, and several months of intermittent uterine bleeding. The patient is a non-smoker with no history of recent surgery, travel, or family history of thrombophilia. While in the ED, the patient decompensated with tachycardia, hypertension, and rapid hypoxemic respiratory failure. Suspected severe cardiogenic shock required rapid initiation of extracorporeal membrane oxygenation (ECMO) to treat the suspected large thrombus. Following initiation of ECMO it was determined to repeat the sequence of imaging utilizing an ECG gated triple rule-out computed tomography (TRO-CT) with delayed imaging of the lower chest, abdomen, and pelvis. The TRO-CT detected a large mobile filling defect within the right atrium measuring 41.2 x 77.7 mm. The large mass was also shown to be hyper enhancing on the delayed exam of the lower chest. The mobile mass was not attached to any of the cardiac structures. The mobile mass was also shown to be intermittently obstructing the tricuspid valve on

the cine data sets as it traversed into the right ventricle. There was no evidence for pulmonary emboli. Review of delayed enhanced CT shows a mass in the right atrium that's connected to a tubular mass that arose as an extension of the filling defect within the inferior vena cava and arising from the left parametrial veins via the left gonadal vein. The delayed enhanced CT also demonstrated a fibroid uterus. Given that the ECMO cannula did not appear to remove any of the mass a thrombus diagnosis is ruled out from the differential diagnosis. Therefore, a diagnosis of a benign intravenous leiomyomatosis was considered. A two-stage surgical removal was planned, with first stage sternotomy and removal of the intracardiac thrombus to manage the patient's cardiogenic shock. The ECMO circuit was stopped and the existing femoral venous cannula was converted to the venous cardiopulmonary bypass circuit. A right atriotomy incision was made and the mass was excised from fibrous adhesions to the right atrial free wall and septum. The portion of the mass that was extending across the tricuspid valve into the right ventricle was delivered into the right atrium. Additional connections at the right atrium/inferior vena cava (IVC) junction and along the tumor extension into the IVC were divided with a combination of blunt and sharp dissection. At this point, the mass was pulled up from the vena cava as much as possible and then transected. Findings were as expected based on imaging. A dense mass occupying most of the right atrium with extension into the right ventricle and inferior vena cava (Figure 1A,B,C gross). For the second stage of the surgery, patient was taken for hysterectomy and unilateral salpingo-oophorectomy. A vertical midline incision was utilized for exposure, allowing an infraumbilical incision for the hysterectomy, with extension into the epigastrium for the remainder of the operation. The right ovary and tube were uninvolved and normal in appearance. The uterus contained several leiomyomas, and the left adnexa was irregular with dilated vessels in the adnexa due to the infiltration of the IVL. The left tube and ovary were removed en-bloc with the uterus to mitigate blood loss and ensure complete resection of the IVL (Figure 1D gross). During this time, the vascular surgeon extended the incision from the xiphoid process to the pubis followed by a right medial visceral rotation. Both the renal vein and IVC were exposed and a transverse venotomy was made (Figure 1E,1F gross) and all parts of the proximal tumor were removed. A hepatobiliary surgeon was also involved to mobilize the liver and gain control of the retro hepatic IVC. The tumor was successfully excised, and hemostasis was achieved. Microscopic pathology confirmed the definitive diagnosis of metastasizing leiomyoma. The patient recovered from both surgeries and was discharged in stable condition with no recurrence.

## Discussion

The management of ICL can be challenging, and with the scarcity of case reports in the literature, it is valuable to report clinical methods used to help manage ICL cases. In our patient, ECMO was used as a bridge to treatment ECMO that provided oxygenation and circulatory support resulting in extended survival in our patient. Severe respiratory and cardiac failure support is a Function of ECMO that has been described in the literature [5]. ECMO technology was developed in the 1970s, with the main purpose to serve as a long-term bypass machine for neonates with severe respiratory failure. Moreover, the diagnosis of ICL can be confused with other medical conditions such as intracardiac thrombus and cardiac angiosarcoma on CT imaging ref. TRO-CT imaging used in this case played an integral role in the diagnosis of IVL with cardiac extension. This imaging method identified the ICL masses originating from uterine leiomyomatosis. Due to the enhancing capability and the presence of a free-floating mass within the right atrium, the TRO-CT imaging assisted with differentiating the diagnosis. To achieve treatment in this case, a surgical resection of the tumor was performed. Common strategies for surgical treatment of ICL have been utilized using a one-stage or two-stage manner [2,4]. Due to the instability and the severity of cardiogenic shock, surgeons decided the use of a double-staged surgical technique that involved the removal of intracardiac and abdominal/pelvic components. In terms of safety and ease, a two-stage surgery is often considered by many studies [5,6]. Complete resection of the tumor is curative, in opposition to a partial removal showing a 1/3 recurrence rate [2].

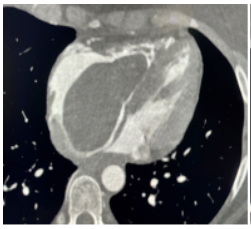

Figure 1A: TRO-CT shows a large low attenuation mobile mass in right atrium which extends into the right ventricle and partially obstructs the tricuspid valve.

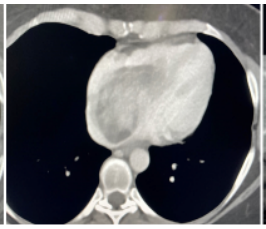

Figure 1B: Delayed venous phase image 2 minutes after ECG gated image in Figure 1A, shows contract enhancement of the large right atrial mass.

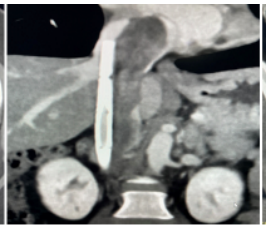

Figure 1C: Coronal post contrast delayed image of the abdomen and pelvis shows the stalk line appearance of a hyper enhancing mass arising from the inferior vena cava with an ECMO cannula also within the inferior vena cava.

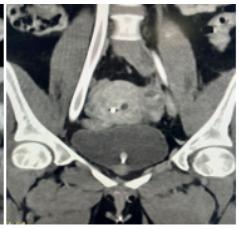

Figure 1D: Coronal reconstruction of the delayed post contrast enhanced exam shows a fibroid uterus with an IUD in place.

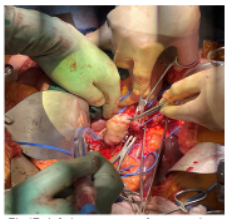

Fig 1E: inferior vena cava after extraction.

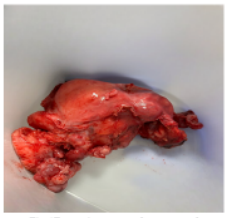

Fig 1F: uterine mass after removal.

### Conclusion

To our knowledge this is the first case to report an ICL presentation with cardiogenic shock that was successfully treated with ECMO and utilized the TRO-CT to assist with diagnosis placing ECMO provided potential beneficial outcomes to the patient in terms of survivability. Findings from this case suggest that using ECMO for bridging patients with ICL presenting with cardiogenic shock is favourable, and further cases using ECMO for complex cases such as the one presented should be reported in the literature.

### Learning Objective

1. To be able to make a differential diagnosis of IL with ECG gated triple rule out CT
2. To understand the role of ECMO utilization in treating IL with cardiac involvement

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