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# Cardiac metastasizing leiomyoma: A case report

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ABSTRACT

*INTRODUCTION:* Cardiac intracavitary growth of metastasizing tumour is unusual. Benign Metastasizing Leiomyoma (BML) from the uterus to the heart is extremely rare. It affects premenopausal women with a history of uterine leiomyoma.

*PRESENTATION OF CASE:* We report a case of a 42-year-old woman who presented three tumours in the right side of the heart, two years after a hysterectomy due to leiomyomatosis. The cardiac tumours were resected and the diagnosis was uterine leiomyoma.

*DISCUSSION:* The patient developed cardiac failure due to three masses at the right side of the heart. Cardiac involvement in BML is usually asymptomatic and rare. The heart masses were surgically removed and a peri ovarian mass was detected and also removed.

Although histologically benign, BML exhibits metastatic qualities.

*CONCLUSION:* It suggests that BML should be included as a differential diagnosis when a female patient presents an intra cardiac mass and a history of hysterectomy.



## 1. Introduction

Although primary cardiac tumours are extremely rare, secondary are not [1]. Secondary metastatic heart tumours occur 20–30 times higher incidence than primary lesions and are uniformly malignant [2]. Despite their frequency, metastatic heart tumours only rarely gain clinical attention because intracavitary growth is unusual [3]. Benign Metastasizing Leiomyoma (BML) is very rare. It was first described 80 years ago [4], and the lung is the most common site for metastasis [5]. We report a case of a cardiac BML that compromised the heart and the pelvis and that was treated at our institution.

## 2. Case report

A 42-year-old female patient presented to the hospital with a worsening retrosternal pain and shortness of breath on moderate efforts that started two weeks before. Her past medical history included hysterectomy for leiomyomatosis two years earlier. As propaedeutic procedure for pelvic pain she had recently performed abdominal ultrasound that showed a left peri ovarian solid mass. Surgery for resection of the mass could not be performed due to the current symptoms of cardiovascular disease.

She had no other significant prior history. She denied smoke, alcohol intake or substance abuse. She had no oncological familiar history.

Pulmonary thromboembolism (PTE) was excluded by computed tomography (CT) angiography. A transthoracic echocardiography performed in the emergency room revealed a large mass located in the right ventricular cavity and apparently adhered to the interventricular septum, right ventricle (RV) enlargement, a probable thickening of the tricuspid sub valvar apparatus, severe tricuspid regurgitation, pulmonary systolic arterial pressure of 35 mmHg and normal left ventricle (LV) dimension and systolic function (Fig. 1). In order to provide additional characterization of the mass a cardiac magnetic resonance imaging (CMR) was performed and confirmed two large and independent masses, both with tissue characterization pattern compatible with tumour and located in the right ventricular cavity. The largest one, measuring 5.3 × 2.7 cm, was adhered to the anterior face of the right ventricular outflow tract (RVOT) and was responsible to a significant RVOT obstruction. The second mass was multilobulated, pedunculated and mobile attached to the interventricular septum. Moderate global systolic deficit (estimated ejection fraction of 39%) was reported, attributed to the RVOT obstruction (Fig. 2).

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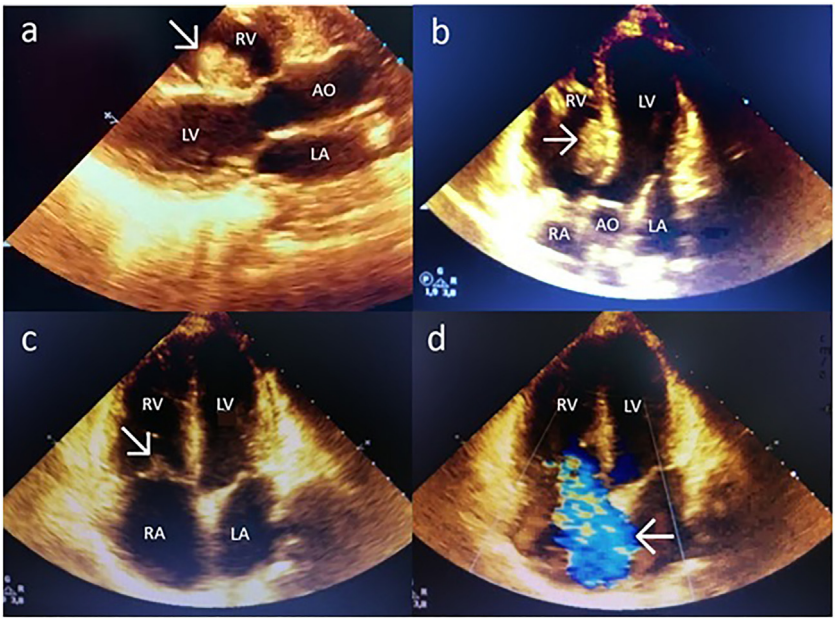


**Fig. 1.** Right Heart Masses on Echocardiography: (a) long axis parasternal view showing a large mass (arrow) adhered to the right side of the interventricular septum; (b) anteriorly angulated four chamber apical view showing the same mass (arrow); (c) four chamber apical view showing right chambers enlargement and thickening of the tricuspid subvalvar apparatus (arrow); (d) severe tricuspid regurgitation (arrow). LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

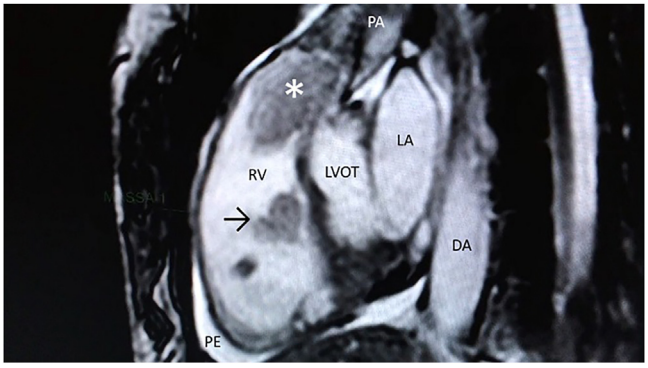


**Fig. 2.** Cardiac magnetic resonance imaging showing a large mass (*) and a smaller one (arrow) inside the RV. DA: descending aorta; LA: left atrium; LVOT: left ventricle outflow tract; PA: pulmonary artery; PE: pericardial effusion; RV: right ventricle.

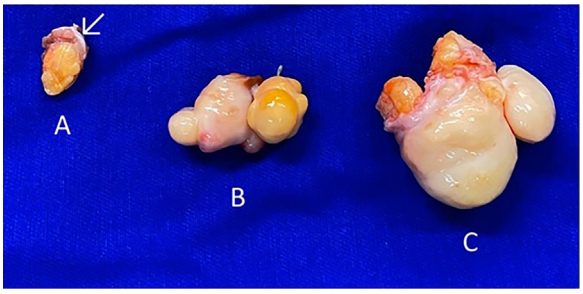


**Fig. 3.** Surgical specimen consisting of 3 pink-tan rubbery masses. A: mass adhered to the tricuspid valve (valve tissue - arrow); B: mass attached to the interventricular septum; C: mass attached to the anterior face of the RVOT.

After the magnetic resonance result, she was submitted to a cardiac surgery with cardiopulmonary bypass for tumours resection. The surgery was performed by median sternotomy, with cannulation of the aorta, superior and inferior vena cava. Anterograde cardioplegia was made. Intraoperatively, a third small neoformation was identified heavily incorporated to the ventricular face of the anterior leaflet and to the chordae of the tricuspid valve (Fig. 3). The masses were completely excised with sacrifice of the tricuspid valve. A valve replacement was mandatory. The surgery was performed with no complications and the immediate postoperative course was uneventful. Furthermore, the postoperative control transthoracic echocardiography showed the normal functioning tricuspid valve, without any obstruction of the RVOT and preserved systolic function of both ventricles (Fig. 4).

Pathology of the cardiac masses revealed findings consistent with a benign smooth muscle neoplasm.

Histologically, the tumour was a mesenchymal neoplasm compatible with leiomyoma, no mitosis, areas of necrosis or signs of malignancy were found. The segment of the cardiac valve in the specimen had normal histological appearance.

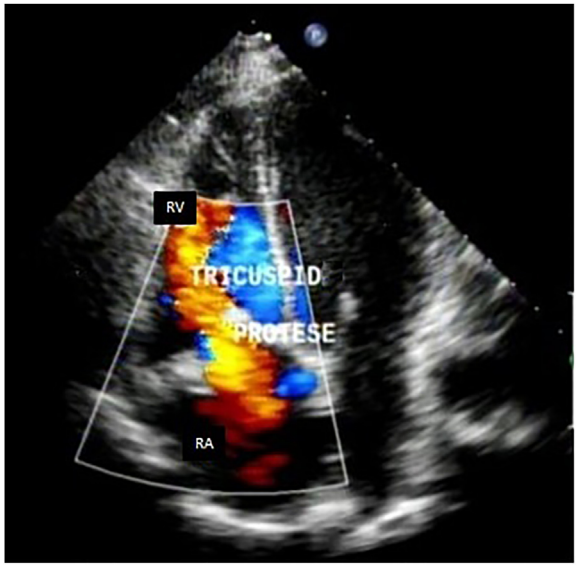


**Fig. 4.** Four chamber apical view showing normal function tricuspid valve prosthesis and RV with systolic function preserved.

Immunohistochemically, tumour cells were positive for smooth muscle desmin and caldesmon; and estrogen, supporting a gynecological leiomyoma lineage.

As the patient had a previous diagnosis of a periovarian solid mass, she was referred to the gynecology team and was submitted to a magnetic resonance imaging of the pelvis. It revealed a large mass next to the left ovarian. She was submitted to a laparotomy nine days after the cardiac surgery and the mass was resected. Microscopic pathology revealed a leiomyoma, without atypia, mitosis or necrosis. She recovered from the second surgery without complications and was discharged from the hospital followed by the gynecology service. The final diagnosis was benign metastasizing leiomyoma to the right ventricle and pelvic.

## 3. Discussion

Metastasis can reach the heart via lymphatic route, hematogenous route, transvenous extension and direct extension [6]. Lymphatic tends to give rise to pericardial and epicardial masses, while hematogenous spread preferentially gives rise to myocardial or endocardial [3]. In general, benign tumours involve the left side of the heart and malignant, the right side.

Most cardiac metastasis are clinically silent and are diagnosed only post-mortem. When they manifest, may imitate valvar heart disease or cause cardiac failure, ventricular or supraventricular heart rhythm disturbances, conduction defects, syncope, embolism, or, quite often, pericardial effusion [3].

The most common tumours with cardiac metastatic potential are carcinoma of the lung, the breast and the esophagus, malignant lymphoma, leukemia and malignant melanoma [3].

Cardiac magnetic resonance (CMR) has become the gold standard for evaluation of such masses and allows optimal tissue differentiation and accurate characterization of the mass preoperatively [6,7]. CMR enables the differentiation of intra cardiac thrombus from a tumour due to avascular tissue composition [8].

We described the case of a pre-menopausal woman who developed cardiac failure due to three masses at the right side of the heart. The histological findings were compatible to benign metastasis of leiomyoma. The patient cardiac tumours were immono-reactive for estrogen, desmin and caldesmon confirming the diagnosis.

Benign metastasizing leiomyoma is an extra uterine smooth muscle tumour that occurs in patients with a current or prior history of uterine leiomyoma [8]. These are the most common gynecological tumours in pre-menopausal woman, found in up to 30% of women older than 35 years [9].

Different types of extra uterine growth of benign uterine leiomyomas are described as follows: disseminated peritoneal leiomyomatosis (DPLM), retroperitoneal leiomyomatosis (RPLM), parasitic leiomyoma, BML, and intravenous leiomyomatosis (IVL) [10]. Despite being histologically benign, leiomyoma has clinically malignant potential. The most frequent site of metastasis is the lungs, although other areas may also be affected as well, including some atypical locations, e.g. the heart or spinal cord [11].

Based on the literature, BML, DPLM and RPLM have been observed to be more indolent [10,11]. BML is a very rare disease that has been reported in association with uterine leiomyoma, and about 100 cases have been reported in the literature [12]. To our knowledge, this is the FOURTH case of BML affecting the tricuspid valve [13], and the TENTH case of heart involvement reported in the literature [13]. The pathogenesis of BML remains unclear. However, it has been postulated BML spreads hematogenously, originates from independent multiple foci, and/or is hormone-driven [10,11].

BML may present many years after the uterine surgery. The mean time from primary surgery to BML diagnosis was 8,8 years according to a recent review [14]. In the case reported the hysterectomy was performed 2 years before the diagnosis of cardiac BML.

Uterine leiomyomatosis is highly hormone sensitive, and treatments are based on hormonal manipulation with either surgical or medical castration [15]. Hormone suppression has been shown to either stabilize or even induce regression of metastatic lesions [16].

Medical treatment alone is highly insufficient in the case of intra cardiac tumours due to the risk of heart failure and possible sudden death caused by total outflow tract obstruction. Regardless of the pathogenesis of intra cardiac leiomyomatosis, review of the literature suggests that surgical removal of the intra cardiac tumor is curative (19). Complete removal is strongly recommended, as no recurrence has been reported with total resection, as opposed to 1/3 recurrence rate in patients who underwent partial resection [17].

The SCARE 2018 was followed to improve the reporting transparency [18].

Based on this case, we suggest that BML should be included as a differential diagnosis when a female patient presents an intra cardiac mass and a history of hysterectomy.

## Declaration of Competing Interest

We have no conflicts of interest.

## Sources of funding

There are no sources of funding for our research.

## Ethical approval

Approval was obtained from the hospital ethics committee.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Consent

Patient privacy was respected all over the paper.

## Author contribution

Every author worked together in the Study conception and design, Acquisition of data, Analysis and interpretation of data, Drafting of manuscript, and Critical revision.

## Registration of research studies

1. Name of the registry: do not apply
2. Unique identifying number or registration ID: do not apply
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): do not apply

## Guarantor

Soares, Raquel Reis.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

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