

Received: 2019.11.02

Accepted: 2020.01.03

Available online: 2020.04.01

Published: 2020.04.29

Leiomyosarcoma Tumor Embolism Masquerading as Thrombus in Transit

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 58-year-old
Final Diagnosis: Tumor embolism
Symptoms: Dyspnea
Medication: —
Clinical Procedure: Percutaneous embolectomy • surgical embolectomy
Specialty: Cardiac surgery • Cardiology • Critical Care Medicine • Oncology

Objective: Rare disease

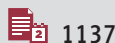
Background: Tumor embolism is a rare neoplastic complication that occurs when there is intravenous invasion by a benign or malignant tumor. We present the case of an asymptomatic patient with an incidentally discovered leiomyosarcoma tumor emboli, which was initially misdiagnosed as “thrombus in transit.”

Case Report: The patient was a 58-year-old woman who was incidentally found on echocardiogram to have a large tubular mass within the inferior vena cava and right atrium. Although initially characterized as “thrombus in transit”, this mobile right atrial mass was present without clinical, echocardiographic, or radiographic evidence of pulmonary embolism or increased pulmonary arterial impedance. Given that a thrombus in transit is nearly always associated with submassive or massive pulmonary emboli and their attendant right heart sequelae, these pertinent negative findings led us to seek an alternative diagnosis. After a trial of conservative management with anticoagulation and attempted removal of the mass with the AngioVac system, the patient ultimately underwent median sternotomy and surgical embolectomy on cardiopulmonary bypass to remove the mass, which was later identified on pathology as a leiomyosarcoma.

Conclusions: With rare exceptions, “thrombus in transit” is accompanied by large pulmonary emboli and the presence of increased pulmonary artery pressure and right heart strain. The absence of clinical, echocardiographic, or radiographic evidence of these hemodynamic sequelae should raise suspicion for an alternative diagnosis. Tumor embolism should be considered in the differential diagnosis of any patient with a history of malignancy who presents with evidence of intracardiac mass or embolism.

MeSH Keywords: Neoplastic Cells, Circulating • Pulmonary Embolism • Ventricular Function, Right

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/921124>



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Background

Tumor emboli are a rare complication of malignancy that typically present with subacute, non-specific symptoms and have a poor prognosis [1]. In most cases described in the literature, patients present with progressive symptoms such as dyspnea, chest pain, palpitations, and lower-extremity swelling [2–5]. Tumor emboli occur when there is intravenous invasion by a benign or malignant tumor. Most commonly associated with renal, breast, lung, or gastric cancers, tumor emboli from leiomyosarcoma are extremely rare, with only a few case reports worldwide [6]. Further, we have found no case reports of leiomyosarcoma tumor emboli incidentally discovered in an asymptomatic patient. We present the case of a patient with an incidentally discovered uterine leiomyosarcoma embolus in the inferior vena cava and right atrium.

Case Report

A 58-year-old woman initially presented to her physician complaining of abdominal pain, uterine bleeding, and a palpable abdominal mass. Her symptoms were attributed to uterine fibroids and she underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Pathology revealed that her uterine mass was in fact a malignant leiomyosarcoma and she was started on doxorubicin-based chemotherapy (CHOP). Shortly after beginning her treatment course, she had a routine transthoracic echocardiogram to screen for chemotherapy-induced cardiomyopathy. The echocardiogram demonstrated normal size and function of the right and left heart and no Doppler evidence of increased pulmonary vascular resistance. The report also noted a “large, mobile, tubular mass entering the right atrium through the inferior vena cava, consistent with thrombus in transit.” The patient was asymptomatic and hemodynamically stable at this time but was admitted to the Cardiac Intensive Care Unit and started on high-dose heparin (aPTT 80–100). The pulmonary hypertension and right heart failure cardiology service was consulted.

Importantly, chest computed tomography angiography was negative for pulmonary embolism and there was no evidence of right heart strain, consistent with the echocardiographic findings. In addition, venous duplex ultrasound of the bilateral lower extremities was negative for deep vein thrombosis. After 1 week of treatment with anticoagulation, a transesophageal echocardiogram showed no decrease in the size of the mass and redemonstrated normal right heart morphology and function (Figure 1). Given the lack of response to heparin, percutaneous removal of the mass was attempted using the AngioVac system (AngioDynamics, Latham, NY) under transesophageal echocardiographic guidance. During the procedure, the mass dislodged and embolized to the right pulmonary

artery, preventing its retrieval with the AngioVac device. That night, the patient became mildly febrile and tachycardic, with a heart rate of 110–120 bpm. The following day, she underwent midline sternotomy and surgical embolectomy on cardiopulmonary bypass. A 14-cm, white, well-organized mass was removed from the right pulmonary artery (Figure 2). Pathology confirmed a tumor consistent with leiomyosarcoma. The patient had an uncomplicated post-operative course and was discharged on post-op day 12 with outpatient oncology follow-up.

Discussion

Herein, we report the case of a patient with a recent diagnosis of leiomyosarcoma who presented with an asymptomatic, mobile, intracardiac mass. Given the predisposition to venous thromboembolism in the setting of active malignancy, as well as the appearance and location of the mass, thrombus in transit associated with pulmonary embolism was high on the differential diagnosis. However, the lack of pulmonary vascular imaging evidence of pulmonary embolic material and the patient’s normal right heart size, shape, and function argued against this diagnosis. This, combined with a lack of response to systemic anticoagulation, led to the alternate diagnosis of tumor embolus. This observation is especially relevant in the era of the pulmonary embolism response team (PERT), where rapid intervention and use of thrombolytics may occur without a definitive diagnosis being made. Given the significantly different management strategy required for tumor emboli, careful consideration must be given to the differential diagnosis before treatment of a caval or right atrial embolism is begun.

Intravenous spread and embolization of uterine leiomyosarcoma to the inferior vena cava, right atrium, and pulmonary artery is extremely rare, and in all previous case reports it was diagnosed in symptomatic patients. In this case, the mass was detected incidentally before it could progress and cause circulatory compromise. Most patients with tumor emboli present with symptoms similar to those with venous thromboembolism such as dyspnea and hypoxia [1–6]. Further, it is well established that active malignancy increases the risk for thrombosis, which may bias physicians towards a diagnosis of venous thromboembolism [7]. Often, the diagnosis of tumor emboli is made only after the failure of anticoagulation for suspected venous thromboembolism [1]. In our case, the right atrial mass did not decrease in size following treatment with high-dose anticoagulation. This suggested that the mass was composed of an alternative, non-thrombotic material. Although difficult to distinguish from thrombotic emboli, correct diagnosis of tumor emboli is critical, as the management differs significantly from that of thrombotic emboli. Lessons extrapolated from the more common and widely studied phenomenon of renal carcinoma tumor emboli dictate that surgical embolectomy on

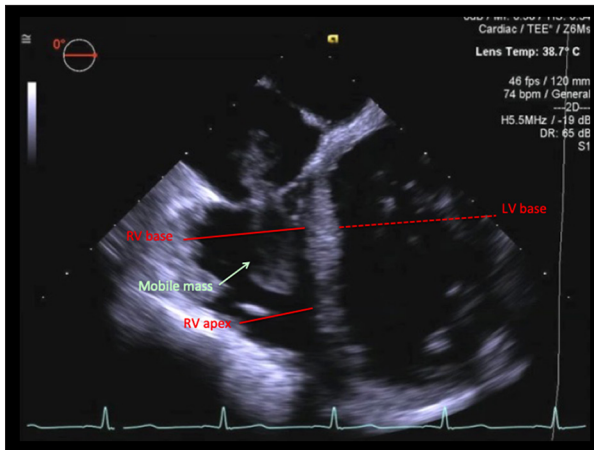


Figure 1. Transesophageal echocardiogram four-chamber view demonstrating normal right ventricle size (RV: LV base dimension ratio <1.0 ; RV solid line and LV dashed line) and morphology (RV base: apex ratio >1.5 ; solid lines), both of which are inconsistent with pulmonary hypertension. Note the large tubular mass crossing the tricuspid valve.

cardiopulmonary bypass is often required when tumor emboli exist in the vena cava and right atrium [8,9].

A mobile, worm-like, right atrial mass most commonly is a thrombus in transit, which is nearly always associated with a large pulmonary embolism (98% incidence of PE) [10]. A study by Chartier et al. showed that in patients with free-floating right heart thrombi, pulmonary embolism was most common, and most patients had evidence of right heart dilatation, dysfunction, and increased pulmonary vascular resistance. In that study, 84% of patients were in New York Heart Association Functional Class IV heart failure and 52% were in cardiogenic shock [11]. A pertinent negative of this case is that the patient was asymptomatic, and despite the initial echocardiographic report of a “thrombus in transit,” there was no evidence of pulmonary embolic material on computed tomography angiography. In addition, transthoracic and transesophageal echocardiograms did not reveal any evidence of right heart dilation, dysfunction, or increased pulmonary vascular impedance. Notably, the right ventricular base: apex diameter ratio was greater than 1.5 and the right ventricle: left ventricle



Figure 2. Photograph of mass surgically removed from patient's right pulmonary artery.

base dimension ratio was less than 1.0; both findings are highly consistent with normal pulmonary arterial impedance and are thus inconsistent with pulmonary vascular obstruction (Figure 1) [12]. Given the nearly complete association of mobile, worm-like right atrial thrombi with pulmonary emboli and their right heart sequelae, these pertinent negative findings raised the suspicion for an alternative, non-thromboembolic mechanism to the right atrial mass [10,11].

Conclusions

In conclusion, a diagnosis of “thrombus in transit” in the absence of clinical, echo-Doppler, or radiographic evidence of pulmonary embolism should be treated with skepticism and an alternative diagnosis sought. This is particularly true when the mass in question is non-responsive acutely to standard anticoagulation therapy. This case highlights the importance of maintaining a high index of suspicion for tumor emboli in patients with a history of malignancy presenting, symptomatic or not, with evidence of intracardiac mass or embolism.

Conflict of interests

None.

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