Cardiac Metastasis of Nonvisceral Soft-tissue Leiomyosarcoma

Cristina Martinez, MD,[1] Jamal S. Rana, MD, PhD,[2-4] Matthew D. Solomon, MD, PhD[2,3]
[1]Department of Emergency Medicine, Alameda Health System - Highland Hospital, Oakland, CA; [2]Department of Cardiology, Kaiser Permanente Oakland Medical Center, Oakland, CA; [3]Division of Research, Kaiser Permanente Northern California, Oakland, CA; [4]Department of Medicine, University of California San Francisco, San Francisco, CA

Metastasis of a nonvisceral leiomyosarcoma to the heart is rare. We present the case of a man with a history of an upper extremity cancerous lesion that was completely resected with appropriate surveillance monitoring, which then metastasized to the heart 14 years later, presenting as superior vena cava syndrome. Full evaluation found no other metastatic lesions, including no residual sarcoma at the former primary site. We include transthoracic echocardiography and computed tomography images of unusual presentation of the large mass extending from the caudal superior vena cava to the right atrium and into the right ventricle across the tricuspid valve.

[Rev Cardiovasc Med. *2017;18(2):78–81 doi: 10.3909/ricm0859*]

KEY WORDS

Cardiac mass • Metastasis • Nonvisceral leiomyosarcoma

Primary cardiac tumors are rare, whereas metastatic tumors are up to 20 times more common.[1,2] Leiomyosarcoma (LMS) is a soft-tissue sarcoma that is characterized by smooth muscle cells. It is found throughout the body, although it often occurs in the uterus. Other common sites include the retroperitoneal space, abdominal cavity, and any vascular structure, because it can grow in any tissue with smooth muscle.[3-5] Although there are many case reports of uterine or vascular LMS metastasizing to the heart, we were unable to find any case reports of sole cardiac metastasis from a nonvisceral lesion. Nonvisceral soft-tissue LMS is rare and accounts for only 10% of all soft-tissue sarcomas in adults.[3] We describe this rare case, and review the varying mechanisms by which cardiac masses can cause symptoms.

Case Report

A previously healthy, nonsmoking 63-year-old man presented to the emergency department with progressive dyspnea on exertion for 1 month. He sought treatment for acute onset of swelling to his face, neck, and arms, and 2 days of progressive orthopnea. He complained of no associated chest pain or palpitations, no leg swelling or change in urination, and no history of heart failure. He denied any fevers or weight loss, and a full review of systems was only significant for a dry cough of 3 months.

Fourteen years prior, the patient had a high-grade LMS on the medial aspect of the left arm that was surgically excised, and treated with internal brachytherapy and external beam radiation therapy. A repeat excision performed 3 months later revealed no residual tumor, and results of surveillance magnetic resonance imaging done 2 years later were negative.

Upon arrival, the patient's vital signs revealed blood pressure of 120/93 mm Hg, heart rate of 115 beats/min, respiratory rate of 22 breaths/min, oxygen saturation of 96% on room air, and body temperature of 36.9°C. Physical examination was remarkable for facial swelling/flushing and jugular venous distension. Heart sounds were distant, and no murmurs, rubs, or gallops were heard. The patient was tachypneic, with an increased effort breathing but with clear lungs. There was no peripheral edema, and the extremities were warm and dry.

His initial electrocardiogram revealed sinus tachycardia at 124 beats/min and low voltage throughout, which was new when compared with a prior electrocardiogram from 1999. Results of a chest radiograph were unremarkable, but a bedside echocardiogram was concerning for a large right ventricular (RV) mass. His blood pressure was stable and he was not hypoxic.

Results of a chest radiograph were unremarkable, but a bedside echocardiogram was concerning for a large right ventricular (RV) mass.

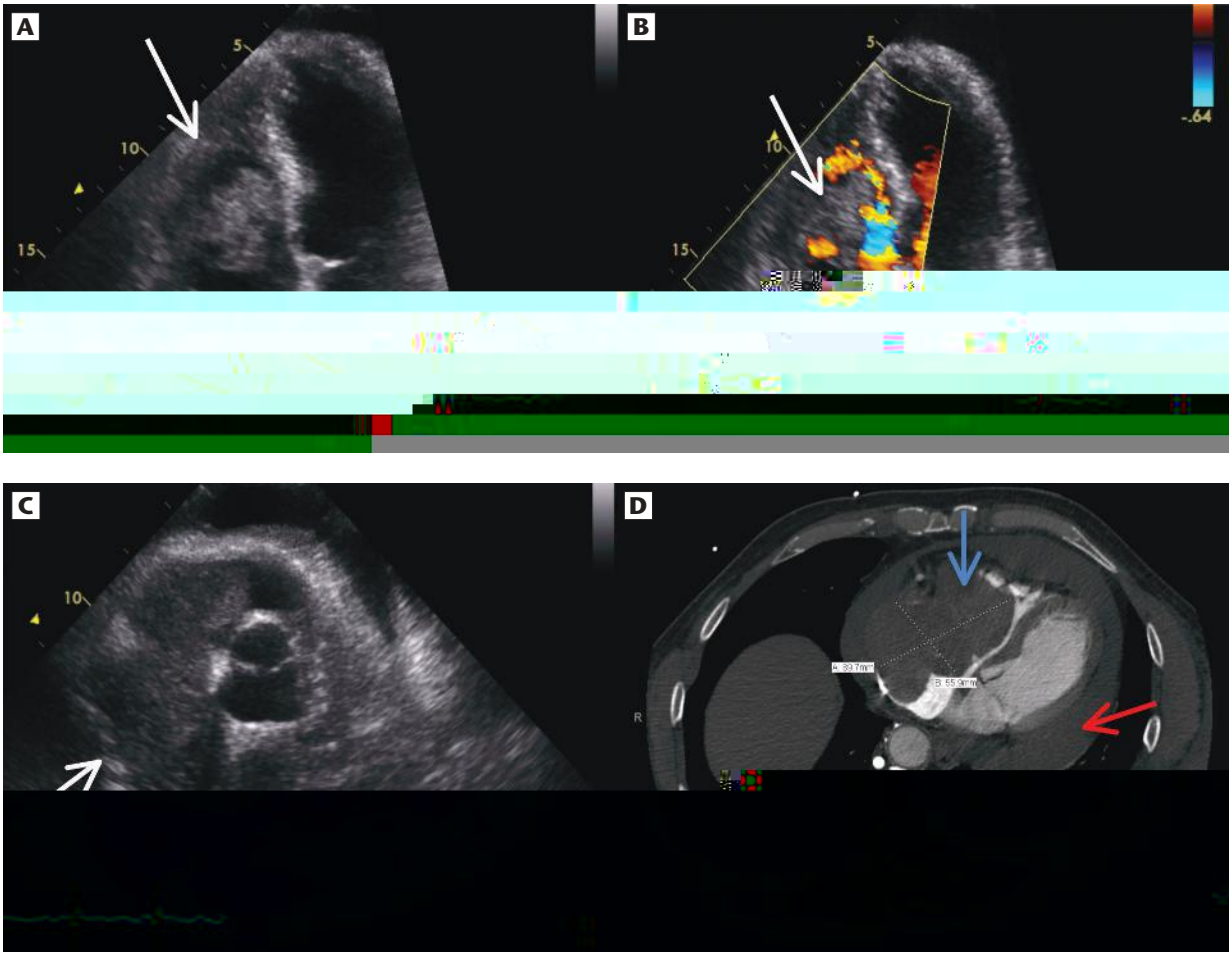

Figure 1. Transthoracic echocardiogram and computed tomographic images of the mass. **(A)** Transthoracic echocardiogram (apical 4-chamber view) showing the large mass in the right ventricle. **(B)** Color Doppler flow surrounds the mass. **(C)** Parasternal short-axis view of the mass invading both the right ventricle and right atrium. **(D)** Computed tomography image of the mass in the right ventricle measuring 89.7 mm × 55.9 mm demonstrated by the *blue arrow* and the large surrounding pericardial effusion demonstrated by the *red arrow*.

A formal echocardiogram showed a large circumferential pericardial effusion and revealed a large intracardiac mass versus thrombus in both the right atrium and ventricle, which measured 9.6 cm in length and 5.2 cm in width (Figure 1). It appeared adherent to the RV free wall, interfered with tricuspid valve function, and obstructed the RV outflow tract. There was also diastolic RV collapse and significant septal flattening of the left ventricle, suggesting high right atrial pressure due to mass effect. There was normal left ventricular size and systolic function, with an estimated ejection fraction of 65%.

A computed tomography (CT) angiogram was performed to characterize the mass and rule out pulmonary emboli. Results of the study showed no pulmonary emboli, but showed the 60 mm × 90 mm mass extensively filling the right heart, extending from the caudal superior vena cava to the atrium, and then into the ventricle across the tricuspid valve (Figure 1). Appearance on CT and echocardiogram was not consistent with a thrombus.

A right heart catheterization was done in order to biopsy the mass and perform therapeutic pericardiocentesis. A total of 580 mL of serous fluid was drained from the pericardial space. Pathology results of the biopsy specimen were reviewed and compared with those taken 14 years ago from his arm. Both showed a high-grade sarcoma consistent with LMS, indicating that this was a metastatic rather than a primary cardiac tumor. A cardiothoracic surgery evaluation was obtained; given that the mass either originated from the RV or had significant invasion into the RV free wall, it was determined it would not be possible to excise the mass and leave sufficient functional RV structure. Full body imaging revealed no other metastatic lesions at that time. Hospice was initiated, in addition to palliative radiation therapy to reduce the tumor size in an attempt to decrease swelling and shortness of breath. The patient expired 5 months later.

Cardiac tumors are often insidious and must always be included in the differential diagnosis of a patient with new cardiac symptoms…

Discussion

Among the unique aspects of this case is the significant delay of 14 years of this LMS of the skin, which had been completely excised with standard care and subsequent surveillance. Although we cannot rule out two primary LMS lesions, it is unlikely because both lesions were high-grade sarcomas consistent with LMS. One of the features used to evaluate the risk of metastasis is the grade of the primary tumor; the high grade of the initial skin lesion put this patient at significant risk for metastasis.[4]

One of the features used to evaluate the risk of metastasis is the grade of the primary tumor…

Another distinct aspect of this case is that the initial metastasis was to the heart alone. Two studies have looked at cardiac metastasis of any soft-tissue sarcoma and both note that it rarely occurs in isolation.[6,7] One of the reasons cardiac lesions are difficult to diagnose as primary metastatic lesions and are often only found incidentally is that other organ systems (such as the lung) often have metastases first, and usually cause noncardiac symptoms that lead to diagnosis.[6-8]

Cardiac tumors are often insidious and must always be included in the differential diagnosis of a patient with new cardiac symptoms such as valve disease, heart failure, arrhythmias, conduction defects, syncope, or pulmonary embolus. Cardiac-related symptoms are often caused by the following mechanisms: valve obstruction leading to a heart failure syndrome; direct invasion of the myocardium, which may cause arrhythmias, embolization of tumor material, intracardiac shunting or pericardial effusion; or constitutional symptoms, which may present similarly to that of an infective, malignant, or immune disease.[1]

Conclusions

Cardiac masses may present heterogeneously as either primary or—more commonly—as metastases from other primary sources. Modern imaging techniques can lead to appropriate and rapid diagnosis. ■

Pathology results of the biopsy specimen were reviewed and compared with those taken 14 years ago from his arm. Both showed a high-grade sarcoma consistent with LMS, indicating that this was a metastatic rather than a primary cardiac tumor.

References

1. Vander Salm TJ. Unusual primary tumors of the heart. *Semin Thorac Cardiovasc Surg*. 2000;12:89-100.
2. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol*. 1996;77:107.
3. Svarvar C, Böhling T, Berlin O, et al; Scandinavian Sarcoma Group Leiomyosarcoma Working Group. Clinical course of nonvisceral soft tissue leiomyosarcoma in 225 patients from the Scandinavian Sarcoma Group. *Cancer*. 2007;109:282-291.
4. Snowden RT, Osborn FD, Wong FS, Sebelik ME. Superficial leiomyosarcoma of the head and neck: case report and review of the literature. *Ear Nose Throat J*. 2001;80:449-453.
5. Rao UN, Finkelstein SD, Jones MW. Comparative immunohistochemical and molecular analysis of uterine and extrauterine leiomyosarcomas. *Mod Pathol*. 1999;12:1001-1009.
6. Hallahan DE, Vogelzang NJ, Borow KM, et al. Cardiac metastases from soft-tissue sarcomas. *J Clin Oncol*. 1986;4:1662-1669.
7. Takenaka S, Hashimoto N, Araki N, et al. Eleven cases of cardiac metastases from soft-tissue sarcomas. *Jpn J Clin Oncol*. 2011;41:514-518.
8. Agaimy A, Rösch J, Weyand M, Strecker T. Primary and metastatic cardiac sarcomas: a 12-year experience at a German heart center. *Int J Clin Exp Pathol*. 2012;5:928-938.

MAIN POINTS

- Cardiac tumors are often insidious and must always be included in the differential diagnosis of a patient with new cardiac symptoms such as valve disease, heart failure, arrhythmias, conduction defects, syncope, or pulmonary embolus.
- One of the reasons cardiac lesions are difficult to diagnose as primary metastatic lesions and are often only found incidentally is that other organ systems often have metastases first, and usually cause noncardiac symptoms that lead to diagnosis.
- Modern imaging techniques can lead to appropriate and rapid diagnosis.
- Leiomyosarcoma (LMS) is a soft-tissue sarcoma that is characterized by smooth muscle cells, and is found throughout the body. Nonvisceral soft-tissue LMS is rare and accounts for only 10% of all soft-tissue sarcomas in adults.