A 34-year-old woman presented to our facility with multiple episodes of hematemesis and left-sided back pain that radiated to her left lower extremity for 3 months. Before her presentation to the hospital, her primary care physician found her to have “a new blowing murmur in the left mid clavicular region fifth intercostal space.” On examination, the patient had a heart rate of 128 beats/min and blood pressure of 125/47 mm Hg. Cardiovascular examination revealed normal S1 and S2 but a grade III/IV diastolic decrescendo murmur best heard at the left precordium midclavicular, followed by a sound consistent with a tumor plop. Results of respiratory, abdominal, and neurologic examinations were unremarkable.

The patient had low serum albumin and total protein, with elevated hemoglobin of 7.7 g/dL. Her tumor markers (carcinoembryonic antigen, cancer antigen 19-9, cancer antigen 125, ß-fetoprotein, and human chorionic gonadotropin) were not elevated; human immunodeficiency virus was nonreactive. Electrocardiography (Figure 1) revealed normal S1 and S2 but a grade III/IV diastolic decrescendo murmur best heard at the left precordium midclavicular, followed by a sound consistent with a tumor plop. Results of respiratory, abdominal, and neurologic examinations were unremarkable.

Computed tomography of the chest/abdomen and pelvis revealed no malignancy. Right pleural fluid aspirate was negative for metastases. Two-dimensional echocardiography revealed a large, irregular, echogenic mass, measuring 46 × 27 mm, located on the anterior leaflet of the mitral valve on the atrial aspect in the left atrium, severe mitral stenosis, mild mitral regurgitation, mild aortic insufficiency, and severe tricuspid regurgitation with an estimated peak pulmonary arterial pressure of 113 mm Hg, with normal left ventricular systolic function and an ejection fraction estimated at 60–65% (Figures 5A and 5B). The parasternal long-axis view demonstrated a large nonmobile mass on the atrial side obstructing mitral inflow (Videos 1 and 2). A short-axis view revealed a mass migrating across the annular plane (Figure 6, Video 3). Also, in the apical four-chamber view, there was evidence of a dilated right ventricle and left atrial mass that appeared adherent to the anterior leaflet of the mitral valve (Videos 4 and 5).

Transesophageal echocardiography (Figures 7A and 7B) confirmed the presence of a large echogenic mass occupying the majority of the left atrial cavity, including a medium-sized spherical echogenic fixed mass on the lateral wall that represented likely tumor metastasis. There was an area of confirmed nonmobile mass associated with the anterior leaflet of the mitral valve (Video 6), and color flow Doppler was visualized circumferential to the mass (Video 7). The left atrial mass was seen extending to the opening of the appendage and moderate bileaflet restriction of the posterior leaflet greater than the anterior leaflet of the mitral valve with severe mitral inflow obstruction. When color flow Doppler was used in the gastric view of the long axis of the left ventricle and mitral valve, it exhibited a mosaic pattern consistent with regurgitation and stenosis (Videos 8 and 9). The mean transmortal gradient was 24 mm Hg with at least mild mitral regurgitation (difficult to quantify) and a large left pleural effusion.

Given the bilateral adrenal lesions seen on computed tomography of the chest/abdomen and pelvis, the patient underwent a right adrenal biopsy. Multiple (more than three) tan-white core biopsies measuring 1.3–1.4 cm were obtained. The core biopsy demonstrated proliferation of atypical spindle cells showing pleomorphic nuclei with nuclear hyperchromasia and occasional mitosis with a myxomatous background: high-grade spindle cell malignancy favoring sarcoma (Figure 8). Further pathology of the duodenum and gastric ulcer revealed no malignancy. Right pleural fluid aspirate was negative for malignancy.
The patient was evaluated by cardiothoracic surgery and oncology, and the mass was determined to be inoperable because of evidence of metastases. Cardiac MRI (Figure 9) confirmed a dominant left atrial mass measuring 7.2 × 3.7 cm, with high T2 signal with partial enhancement compatible with malignancy, mitral stenosis, distribution of the tumor suggestive of primary malignancy (such as angiosarcoma), and finally evidence of metastatic disease with six or seven additional smaller T2 hyperintense lesions in the right and left ventricular walls.

The patient was offered palliative chemotherapy but declined. She died after 3–4 months in hospice care and 7 months after her initial diagnosis.

**DISCUSSION**

Primary cardiac tumors are rare, with seven cases in >12,000 autopsies.\(^1\)\(^\text{,2}\) Seventy-five percent of primary cardiac tumors are benign, with close to half being myxomas; 25% are malignant, with 95% reported as sarcoma.\(^1\)\(^\text{,2}\) The most common sarcoma is angiosarcoma (34%) and undifferentiated sarcoma (24%).\(^1\)\(^\text{,2}\) Spindle cell sarcoma, also known as intimal sarcoma, is one of the rarest primary cardiac malignancies, with a limited number of reported cases.\(^1\)\(^\text{,3}\)\(^\text{,5}\) Symptoms may include obstruction, local invasion, and embolism(s); the first manifestation may be sudden cardiac death.\(^1\) It is more commonly encountered in large arterial blood vessels, affecting the pulmonary...
trunk more than the aorta. The tumor usually infiltrates the left atrium, as seen in our patient. Lung metastases (40%) and extrathoracic metastasis (20%) to kidneys, lymph nodes, brain, and skin are often seen. Tumor emboli are common, causing distant metastases involving bone, peritoneum, liver, and mesenteric lymph nodes. Therefore, this is a highly aggressive tumor that can metastasize quickly. At the time of diagnosis, up to 80% of spindle cell sarcomas have evidence of metastasis.

The differential diagnosis includes angiosarcoma or synovial sarcoma. From a histopathologic perspective, spindle cell sarcoma usually demonstrates positive immunoreactivity for vimentin, desmin, osteopontin, and MDM2. Overall, samples may exhibit large myxoid areas and/or epithelioid appearance of tumor cells.

The diagnosis can be made through imaging modalities such as transthoracic and transesophageal echocardiography. According to American Society of Echocardiography guidelines, complete transthoracic echocardiography is recommended in all patients suspected of having cardiac tumors. However, transesophageal echocardiography may be superior to transthoracic echocardiography in evaluating cardiac tumors, especially myxomas and papillary fibroelastomas. Echocardiography is recommended for surveillance after surgical removal of cardiac tumors that have a potential for high recurrence, such as myomas. Also, the use of contrast with microbubble agents (such as perflutren) may help differentiate vascular tumors from avascular masses such as vegetations and thrombi.

Management of spindle cell sarcoma is not well defined; there are no evidence-based guidelines, with most data acquired from patient experiences. The role of chemotherapy or radiotherapy in the treatment of primary cardiac sarcoma has not proved beneficial. However, there have been selected reports of doxorubicin

Figure 3  Computed tomography of the abdomen: bilateral adrenal metastases (red arrows).

Figure 4  MRI of the lumbar spine: sagittal T1 precontrast on the left (white arrow), on the right (red arrow) postcontrast enhancement of L1, L4, and L5, consistent with metastatic disease.

Figure 5  Two-dimensional echocardiography: parasternal long-axis view and measurement of the left atrial mass (A) (red arrow) and still image of mitral regurgitation circumferential to mass by color Doppler (B).

Figure 6  Short-axis view and mass migrating across the annular plane (red arrow).
demonstrating some improvement in survival. \(^9,10\) Radiotherapy has been used for treatment of positive margins after resections as well as palliation of aggressive localized disease and local recurrences. \(^1\) Resection with negative borders is an ideal option but has been effective in <50% of cases; incomplete resection can lead to recurrence. Whenever possible, complete surgical resection when the tumor is confined to the heart and no metastases in combination with chemotherapy is the best treatment for primary cardiac tumors. \(^5,11\) Heart transplantation may be an option for patients with inoperable sarcomas when there is no evidence of metastases; however, it has been associated with recurrence of malignancy and new tumors stimulated by immunosuppression. \(^11\) The malignancy may remain silent until an advanced stage. There is poor prognosis, with mean survival of 3 months to 1 year; there was a reported case of survival up to 11 years, but the patient did not have widespread metastases. \(^7\)

**CONCLUSION**

Primary cardiac sarcoma is difficult to treat because it may remain silent until advanced stage, and its course is aggressive. Echocardiography is usually the initial imaging modality given that it is noninvasive and widely available. Computed tomography and cardiovascular MRI provide complementary data regarding tumor extent and metastases that influence management.

**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.03.001.

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