Left Atrial Spindle Cell Sarcoma in a 34-Year-Old Woman

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INTRODUCTION

We report the case of a 34-year-old woman who presented with multiple episodes of hematemesis and left-sided back pain that radiated to her left lower extremity for 3 months. During her diagnostic workup, she was found to have a large, nonmobile mass on the left atrial side affecting mitral valve inflow diagnosed by transthoracic echocardiography, transesophageal echocardiography, and cardiac magnetic resonance imaging (MRI); pathologic examination confirmed high-grade spindle cell malignancy favoring sarcoma. The case is unique in that although there have been advances in imaging techniques, early detection of this tumor is limited because of its varied presentation of symptoms and delay in patient presentation.

CASE PRESENTATION

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. She also noted an unintentional 25-lb weight loss over 1 month. 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 total bilirubin of 1.1 mg/dL, but the results of the remainder of her liver function and coagulation studies were normal. She was anemic, with hemoglobin of 7.7 g/dL. Her tumor markers (carcinoembryonic antigen, cancer antigen 19-9, cancer antigen 125, α-fetoprotein, and human chorionic gonadotropin) were not elevated; human immunodeficiency virus was nonreactive. Electrocardiography (Figure 1) revealed sinus tachycardia and a short PR interval, with no evidence of preexcitation and prominent rightward forces.

Computed tomography of the chest/abdomen and pelvis revealed a large, ill-defined mediastinal mass between the thoracic aorta and left atrium with mass effect onto the pulmonary veins and arteries, with findings compatible with disseminated malignancy (bilateral adrenal lesions, hypoenhancement in left hepatic lobe, and mass lesion posterior to the left iliopsoas muscle; Figures 2A, 2B, and 3).

The patient underwent MRI of the lumbar spine and thoracic spine, which revealed multiple enhancing lesions within the T4, L1, L3, and L4 vertebral bodies as well as L5 to S1; these findings were suspicious for metastatic spread (Figure 4).

MRI of the brain 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 transmitral 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.

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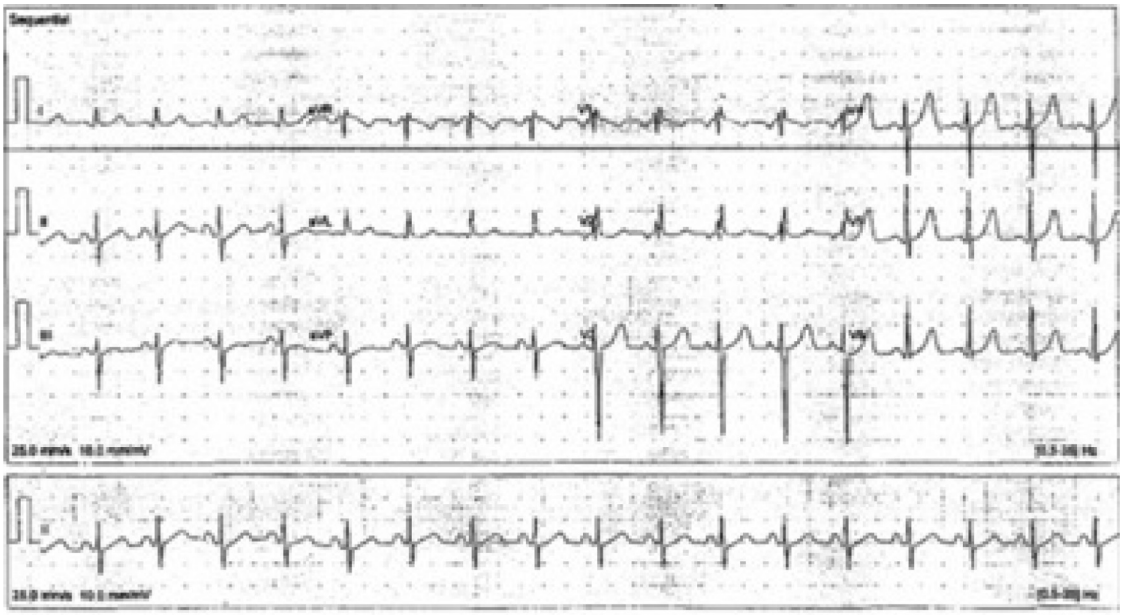
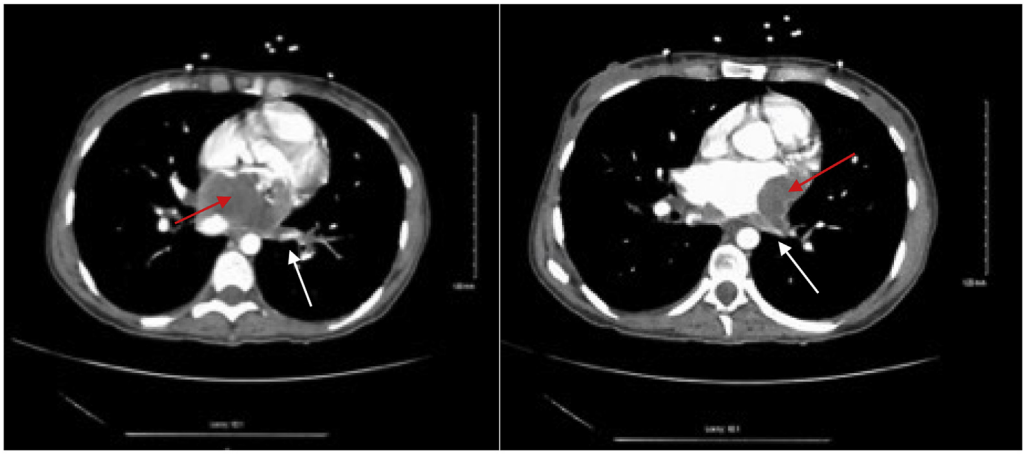


Figure 1 Electrocardiography reveals sinus tachycardia and a short PR interval with no evidence of preexcitation with prominent rightward forces.



a b

Figure 2 Computed tomography of the chest: a large mass **(A)** (*red arrow*), near complete obliteration of the left atrium with impingement of the pulmonary veins **(B)** (*white arrow*).

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.[1,2] Seventy-five percent of primary cardiac tumors are benign, with close to half being myxomas; 25% are malignant, with 95% reported as sarcoma.[1,2] The most common sarcoma is angiosarcoma (34%) and undifferentiated sarcoma (24%).[1,2] Spindle cell sarcoma, also known as intimal sarcoma, is one of the rarest primary cardiac malignancies, with a limited number of reported cases.[1,3-8] 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