

Role of Multimodal Cardiac Imaging

in Diagnosing a Primary Intimal Sarcoma of the Left Atrial Appendage

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Primary cardiac tumors are rare, and most are benign. Intimal sarcomas are among the rarest of the malignant cardiac tumors; they are aggressive and associated with a poor prognosis. Whereas transesophageal echocardiography has been instrumental in evaluating cardiac masses, other imaging methods, such as cardiac magnetic resonance, have proved invaluable in accurately characterizing these masses. We present the case of a 49-year-old woman in whom we diagnosed a primary intimal sarcoma of the left atrial appendage, and we discuss the importance of multimodal imaging in the evaluation and diagnosis of cardiac masses. (Tex Heart Inst J 2019;46(1):28-31)

The frequency of primary cardiac tumors ranges from 0.0017% to 0.25% in autopsy series.¹ Most (75%) are benign, and the most common type is atrial myxoma.¹ Malignant tumors constitute 25% of all cardiac tumors and are predominantly metastatic cancers to the heart.² Most malignant cardiac tumors are sarcomas, which include, in decreasing order of occurrence, angiosarcomas, rhabdomyosarcomas, mesotheliomas, fibrosarcomas and undifferentiated sarcomas, and leiomyosarcomas.³ Cardiac intimal sarcomas are classified as a type of undifferentiated sarcoma, and they are extremely rare. We present the case of a patient who had an intimal sarcoma, and we describe the imaging methods used in the diagnosis.

Case Report

A 49-year-old woman presented to her primary care physician after several weeks of unexplained fever, malaise, and generalized aches. Routine laboratory tests were negative for infection. She was started on nonsteroidal anti-inflammatory medications, which intermittently controlled her symptoms. Her medical history was otherwise unremarkable. She was physically active, and she did not smoke cigarettes, drink alcohol, or use illicit drugs.

The patient continued to have fevers of up to 103° F, so she returned to her physician for further evaluation. Her blood cultures were negative, but a transthoracic echocardiogram (TTE) showed a mass, which was thought to be a vegetation, on the anterior mitral valve leaflet.

On presentation at our institution, the patient was afebrile and hemodynamically stable. The only important finding on physical examination was a grade 2/6 holosystolic murmur, heard loudest at the apex. Her electrocardiogram was normal. She had no peripheral stigmata of infective endocarditis. A transesophageal echocardiogram (TEE) revealed an irregularly shaped, 2.7 × 4.3-cm echogenic mass with fixed and mobile components, originating from and completely filling the left atrial appendage (LAA). The mass extended onto the anterior mitral leaflet (AMVL) and prolapsed through the mitral orifice during diastole (Fig. 1).

Left-sided heart catheterization revealed mild nonobstructive coronary artery disease and the left atrial mass, with vascular supply from the left main and proximal left anterior descending coronary arteries. Because of the size of the mass and its extension onto the mitral valve, as well as the risk for embolization, surgical removal was indicated. Therefore, cardiac magnetic resonance (CMR) was performed to obtain detailed anatomic information for our surgical colleagues. The delayed-enhancement images revealed a heterogeneous mass with areas of thrombus and soft tissue (Fig. 2). There was no evidence of extension of the mass into the myocardium.

Key words: Diagnostic imaging; heart neoplasms/diagnosis/surgery; neoplasm invasiveness; sarcoma/diagnostic imaging/pathology; treatment failure

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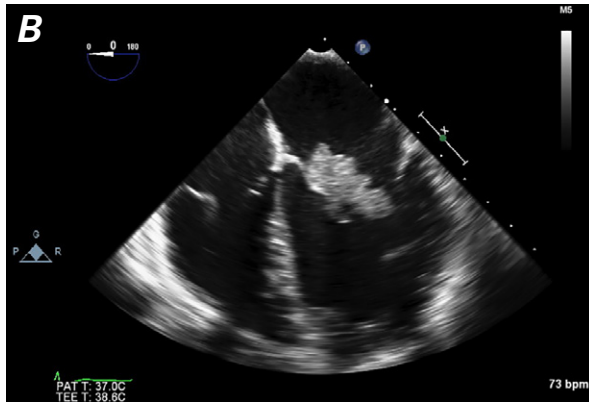
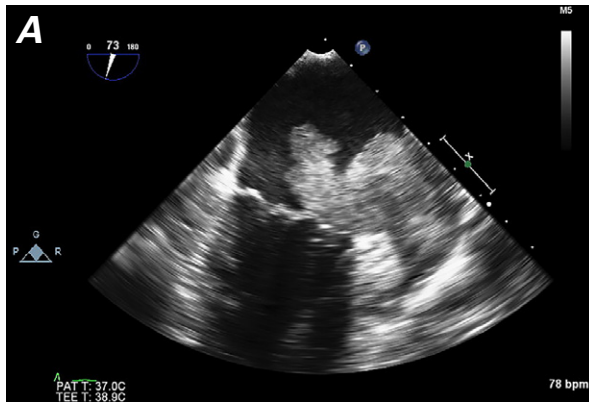


Fig. 1 Two-dimensional transesophageal echocardiograms (midesophageal views). **A)** A large, mobile mass fills the entire left atrial appendage and extends onto the anterior mitral valve leaflet (view at 70°). **B)** The mass is attached to the anterior mitral valve leaflet and extends through the mitral orifice in diastole (view at 0°).

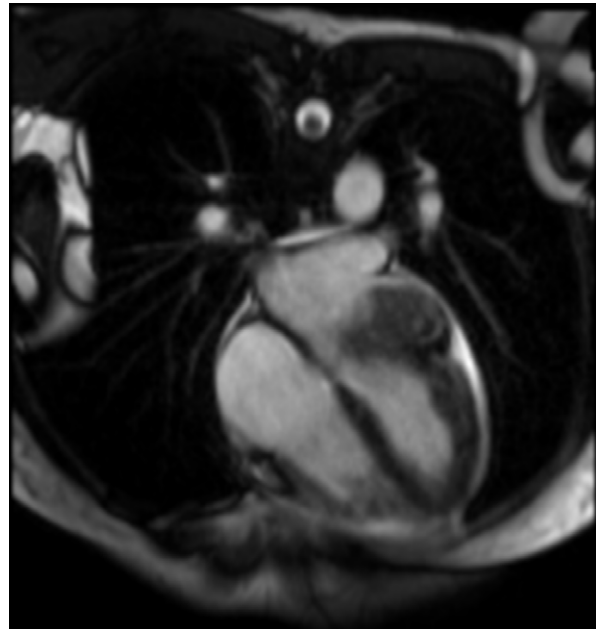


Fig. 2 Cardiac magnetic resonance T2-weighted image (4-chamber view) shows the left atrial mass extending into the left atrial appendage.

At operation, a 5.5 × 3.5 × 2-cm multilobular mass originating from the LAA and extending to the AMVL was resected (Fig. 3). The procedure was challenging because the tumor had to be carefully excised from the AMVL to avoid damaging the annular structures. A postoperative echocardiogram revealed only mild mitral regurgitation.

Microscopic evaluation of frozen sections further characterized the mass as heterogeneous with spindle, epithelioid, and rhabdomyoblast cells, and there was extensive necrosis (Fig. 4). Results of immunohistochemical staining for murine double minute 2 (MDM2), desmin, and focal myogenin were positive. In situ hybridization revealed *MDM2* gene amplification in most of the cells. This relatively specific finding, along with the clinicopathologic features, supported the diagnosis of a high-grade intimal sarcoma.

The patient recovered without surgical complications. On postoperative day 5, computed tomograms (CT) of her chest and abdomen revealed several lesions suspicious for metastatic disease, including a right 4th lateral rib lesion, a 4.5-cm liver mass, a 1.2-cm posterior gastric mass, and a 1.3-cm small-bowel mass. Approxi-



Fig. 3 Photographs show **A)** the gross specimen of the multilobular, irregularly shaped atrial mass, and **B)** its cut surface, revealing a yellow, partially gelatinous tumor, with areas of hemorrhage.

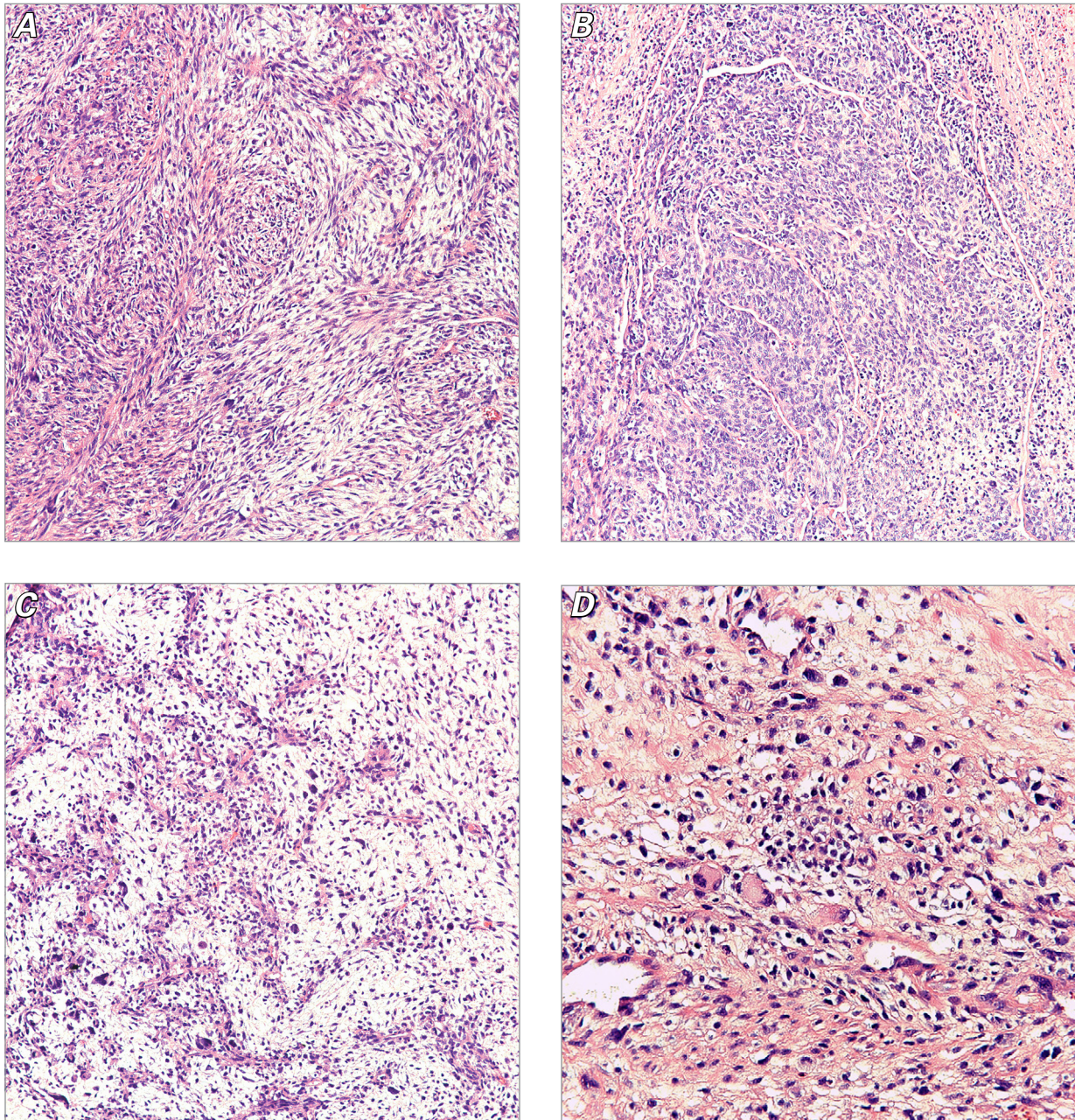


Fig. 4 Photomicrographs show histologic features of the intimal sarcoma: **A)** areas of spindle cells with fascicular growth, **B)** primitive epithelioid cells, **C)** myxofibrosarcoma-like patterns (all H & E, orig. $\times 100$), and **D)** rhabdomyoblastic differentiation (H & E, orig. $\times 200$).

mately one month later, the patient returned to the hospital with nausea, vomiting, and abdominal pain. She was found to have a jejunal intussusception secondary to the small-bowel mass, for which she underwent a minilaparotomy and small-bowel resection. Pathology studies confirmed that the 3-cm mass was a high-grade sarcoma, consistent with metastasis from the atrial sarcoma. Chemotherapy with paclitaxel and gemcitabine was initiated.

Three months later, metastases to the patient's brain were discovered, and she underwent radiation therapy. The following month, repeat CT scans showed interval

progression of metastatic disease involving her brain, liver, and ribs, in addition to recurrence of the LAA mass. Her chemotherapy regimen was changed to gemcitabine, doxorubicin, and vinorelbine. However, her mental status progressively declined, and 8 months after her initial diagnosis, she was transferred to hospice care.

Discussion

Intimal sarcomas are extremely rare mesenchymal tumors that usually arise from the intima of large arteries, most often in the pulmonary trunk or pulmonary arter-

ies, but they can also be found in the aorta.⁴⁻⁶ However, intimal sarcomas that arise from the left atrium, as in our patient, have been reported infrequently.⁷⁻¹²

Our patient's case illustrates the value of multimodal imaging in evaluating cardiac masses. The symptoms caused by cardiac tumors depend on their size, location, and degree of outflow obstruction; they often include dyspnea, chest discomfort, pericardial effusion, tamponade, and heart failure. Our patient had none of these. Her symptoms, which included fever, generalized aches, and malaise, were nonspecific and suggested infection. The initial TTE, however, suggested a valvular mass. Further characterization of the mass through left-sided heart catheterization, TEE, and CMR enabled us to exclude a diagnosis of thrombus or endocarditis and raised concern for malignancy because of its vascular supply, large size, and morphologic appearance. Intimal sarcomas are usually immunoreactive for osteopontin, vimentin, and MDM2.¹³ Our patient's tumor was positive for MDM2.

Intimal sarcomas are aggressive and have a poor prognosis.¹⁴ The mean survival period after diagnosis is 6 to 12 months; however, survival of 11 years has been reported.⁸ Metastatic disease often occurs early in the clinical course, as it did in our patient. Recurrence within several months to a year is typical, despite negative margins during excision.¹⁴ Surgical resection remains the treatment of choice, with consideration of radiation and chemotherapy as adjunctive palliative therapy. However, evidence supporting the effectiveness of chemotherapy and radiation therapy for intimal sarcomas is limited.

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