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Primary Cardiac Sarcoma with Mitral Valve Invasion Complicated by Immune Thrombocytopenic Purpura: A Surgical Dilemma

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Case Report

Keywords: primary undifferentiated cardiac sarcoma, mitral valve involvement, immune thrombocytopenic purpura

Posted Date: November 11th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-10630233/v1

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Additional Declarations: No competing interests reported.

Abstract

Background

Primary cardiac tumors are rare, comprising approximately 0.3–0.7% of cardiac tumors in autopsy series, and sarcomas are the most common primary cardiac malignancy. Most arise in the right atrium as angiosarcoma; left atrial undifferentiated high-grade sarcoma is distinctly uncommon and carries a poor prognosis with low rates of margin-negative resection. Concurrent immune thrombocytopenic purpura (ITP) introduces a management conflict between hemodynamic urgency and prohibitive bleeding risk for which guidance is limited.

Case presentation

A 63-year-old retired firefighter who participated in World Trade Center (WTC) rescue and recovery operations presented with decompensated heart failure. Imaging demonstrated a mobile left atrial mass prolapsing through the mitral valve with severe functional mitral stenosis (mean gradient 15 mmHg). Concurrently diagnosed ITP, confirmed by bone marrow biopsy, produced refractory severe thrombocytopenia (nadir platelet count 24×10³/μL) that repeatedly delayed operative intervention while the lesion was considered possibly thrombotic. After intravenous immunoglobulin and platelet transfusion raised the count to 60×10³/μL, urgent left atriotomy with tumor debulking and bioprosthetic mitral valve replacement was performed. Pathology showed a 6.2-cm high-grade spindle cell malignancy with 28 mitoses per 10 high-power fields, MIB-1 index approximately 50%, absent lineage-specific markers, and a positive posterior left atrial margin, consistent with primary undifferentiated cardiac sarcoma. He recovered and received adjuvant gemcitabine–docetaxel, but developed cerebral, osseous, pulmonary, and diffuse intra-abdominal metastases by 5–6 months and died shortly thereafter.

Conclusions

This case demonstrates how a competing hematologic diagnosis can delay resection of an obstructing cardiac malignancy, and how a microscopically positive margin portends rapid progression in undifferentiated cardiac sarcoma. The temporal relationship between the new ITP and the subsequent tumor diagnosis raises the possibility of a paraneoplastic mechanism, which has been described in roughly a quarter of reported cases of ITP associated with solid tumors as an antecedent finding. The patient's WTC exposure history is documented for completeness; malignancies in this population characteristically present at long latency, but no association between WTC exposure and primary cardiac sarcoma has been established, and attribution is not possible from a single case.

Background

Primary cardiac tumors are rare, accounting for approximately 0.3–0.7% of all cardiac tumors in autopsy series, whereas cardiac metastases are reported to be up to 30 times more common. Of primary cardiac malignancies, sarcomas are the most frequent and typically arise in the right atrium, where angiosarcoma is the predominant histologic subtype. In contrast, left atrial sarcomas are less common, are often initially less infiltrative, and metastasize later.[1] Our patient was found to have an undifferentiated high-grade left atrial sarcoma with direct infiltration into the mitral valve apparatus. Complete surgical resection remains the standard of care; however, overall prognosis for primary cardiac sarcoma remains poor despite aggressive multimodal therapy.[2] Undifferentiated sarcoma accounts for less than 24% of primary cardiac sarcomas and is associated with particularly poor survival and low rates of margin-negative (R0) resection.[3,4] Informed consent for publication was obtained from the patient's family.

Case Presentation

An active 63-year-old, retired firefighter who participated in the World Trade Center rescue and recovery efforts, with cardiometabolic disease and chronic liver disease was emergently transferred from an outside hospital for surgical evaluation of a large left atrial mass. He initially presented with generalized weakness and hypotension, and workup at the referring facility revealed decompensated heart failure with cardiomegaly, pulmonary infiltrates, bilateral pleural effusions, and a 4.1 × 3.5 cm mass along the left atrial wall partially obstructing the left atrial outflow tract on computed tomography angio (CTA) (Figure 1). Transthoracic echocardiogram (TTE) demonstrated a highly mobile left atrial mass (4.8 × 2.2 cm) prolapsing through the mitral valve, causing severe mitral stenosis (mean gradient 15 mmHg) with mild regurgitation (Videos 1 and 2), prompting urgent transfer to our center.

Seven weeks before presentation to our institution, a screening CT of the chest and abdomen obtained for hepatic steatosis follow-up incidentally revealed a low-density mass in the left atrium adjacent to the mitral valve. Cardiac magnetic resonance imaging (MRI) subsequently confirmed a large lobulated left atrial mass prolapsing into the mitral annulus during diastole, and TTE at that time showed a mass prolapsing across the mitral valve causing severe stenosis. Within the same timeframe, he was found to have new-onset thrombocytopenia and, on bone marrow biopsy, was diagnosed with immune thrombocytopenic purpura (ITP). Despite intensive medical therapy, his platelet counts remained severely depressed, repeatedly delaying surgery because of concern for prohibitive bleeding risk and the possibility that the lesion represented thrombus amenable to non-operative management.

On arrival to our institution, he was hemodynamically stable but tachycardic with an irregular rhythm and symptoms consistent with heart failure, and laboratory testing confirmed persistent severe thrombocytopenia in the setting of known ITP (platelet count $24\times10^3/\mu L$). The combination of a highly mobile left atrial mass intermittently obstructing mitral inflow and profound thrombocytopenia posed a unique operative dilemma, balancing the risks of sudden circulatory collapse against prohibitive bleeding on cardiopulmonary bypass. After intensive optimization with intravenous immunoglobulin and platelet

transfusions, his platelet count rose to 60×10³/μL, at which point a multidisciplinary team deemed surgery acceptable, and urgent resection of the left atrial mass was planned.

After median sternotomy and initiation of cardiopulmonary bypass, the interatrial groove was developed and a left atriotomy created. On opening the atrium, a very large mass nearly obstructing the mitral valve was encountered, arising from an approximately 2-cm area of posterior left atrial wall and involving the P1 segment of the posterior leaflet with tumor intertwined in the chordae. The atriotomy was extended behind the inferior vena cava toward the dome of the atrium to accommodate the tumor, and sharp dissection was used to free and debulk the mass, excising it flush with the left atrial wall and leaving a thinned posterior surface highly suspicious for transmural invasion.

Given the extent of involvement, frozen section confirming sarcoma prompted intraoperative discussion with oncology favoring maximal cytoreduction, and the mitral valve was sacrificed to that end. An incision was made along the anterior mitral leaflet, which was excised along with its chordae, as well as the portion of the posterior leaflet adherent to the tumor. Pledgeted horizontal mattress sutures were placed circumferentially around the annulus; in the region of the P1 resection, these sutures necessarily passed through residual tumor. The mitral valve was replaced with a 27 mm bioprosthesis, the left atrial appendage was excluded with an epicardial clip, and the left anterior descending artery was bypassed with the left internal mammary artery. Complete resection was ultimately precluded not by the valve, which was sacrificed, but by transmural tumor invasion of the posterior left atrial wall and by annular extension at P1, with ongoing coagulopathic bleeding on cardiopulmonary bypass (estimated blood loss 1 L, requiring platelets, fresh frozen plasma, cryoprecipitate, packed red cells, prothrombin complex concentrate, and intraoperative immunoglobulin).

Gross examination revealed a 6.2 × 4.5 × 3.0 cm pale tan, nodular left atrial mass with variegated solid cut surfaces, multifocal hemorrhage, and extensive necrosis, along with separate tumor fragments involving the mitral valve apparatus. Histologically, the lesion was a hypercellular high-grade spindle cell malignancy with focal epithelioid areas, myxoid stroma, extensive necrosis, and brisk mitotic activity (up to 28 mitoses per 10 high-power fields). Immunohistochemistry showed weak AE1/3 and MOC31 staining with a proliferation index of approximately 50% by MIB-1, and was negative for muscle, endothelial, mesothelial, melanocytic, and other lineage markers; resection margins were involved along the posterior left atrial wall. On external consultation, these findings were interpreted as a primary high-grade undifferentiated cardiac sarcoma of the left atrium with mitral valve involvement.

Postoperatively, his course was complicated by atrial fibrillation, but he recovered well and was discharged home on postoperative day 8. Staging CT of the chest, abdomen, and pelvis and brain MRI obtained approximately 1 month after surgery showed no definite metastatic disease. About 2 months postoperatively, cycle 1 of 6 of systemic chemotherapy with gemcitabine and docetaxel was initiated for high-grade undifferentiated cardiac sarcoma with a positive posterior left atrial margin. At 5–6 months after surgery, after cycle 4 of chemotherapy, surveillance brain MRI revealed new metastases in the right cerebellum and left occipital lobe and repeat CT of the chest demonstrated a new lytic left fourth-rib

lesion, suspicious pericardial soft tissue, and a new tubular nodular branching opacity in the anterior right upper lobe concerning for metastatic disease. Follow-up CT of the abdomen and pelvis showed rapid progression to extensive metastatic disease involving both adrenal glands, retroperitoneum, perinephric spaces, iliopsoas muscles, and peritoneum, prompting a shift in focus to palliative brain radiotherapy and symptom-directed care. He died 6 days after his follow-up CT scans.

Discussion and Conclusions

This case illustrates the complex interplay between a highly aggressive cardiac sarcoma and severe ITP. New ITP at the time of tumor discovery led to substantial delays in surgical resection as clinicians attempted hematologic optimization, during which the mass progressed to severe functional mitral stenosis.

The sequence of diagnoses raises the possibility of a paraneoplastic mechanism. In a critical analysis of 68 published cases of autoimmune thrombocytopenia associated with solid tumors, ITP occurred concurrently with the cancer in about half of patients and preceded the diagnosis in approximately 25%; most responded to corticosteroids, and durable remission attributable to tumor resection alone was exceptional.[5] Whether this patient's ITP was paraneoplastic cannot be determined retrospectively. The practical implication is that severe thrombocytopenia discovered alongside an intracardiac mass should not be assumed to represent an independent comorbidity, nor should it by itself justify indefinite deferral of resection.

Margin status is the dominant modifiable determinant of outcome and contextualizes his rapid decline. In the National Cancer Database analysis of primary cardiac malignancies, patients with positive margins had significantly reduced long-term survival compared with R0 resection ($p<0.0001$), with overall 1- and 5-year survival of 45.3% and 11.5%.[6] Complete resection was not achievable here because of transmural invasion of the posterior left atrial wall and tumor extension to the P1 region of the annulus; the mitral valve itself was sacrificed in pursuit of maximal clearance. The more radical operations that might have achieved an R0 margin, namely full-thickness posterior atrial wall resection with patch reconstruction or cardiac explantation and autotransplantation, carry substantially longer bypass and cross-clamp times that were not tolerable in a patient already bleeding heavily from refractory thrombocytopenia. Severe ITP therefore constrained not only the timing of surgery but its oncologic completeness. Progression to cerebral, osseous, and diffuse intra-abdominal metastases within 6 months is consistent with the natural history after an R1 resection of high-grade disease.

His history as a firefighter and 9/11 responder is notable. In a pooled cohort of 57,402 World Trade Center rescue and recovery workers followed for 14 years, overall first-primary cancer incidence was below expectation, but incidence of melanoma, prostate, thyroid, and tonsil cancer was significantly elevated, with an exposure–response gradient by arrival date.[7] Malignancies in this population characteristically emerge more than a decade after exposure, and the interval in this patient is consistent

with that pattern. However, no association between World Trade Center exposure and primary cardiac sarcoma has been reported, and causal attribution is not possible from a single case.

Ultimately, surgery proceeded at a platelet count of $60\times10^3/\mu L$, highlighting the need for individualized, multidisciplinary decisions when hemodynamic and hematologic risks are in direct conflict, and further study into optimal surgical, systemic, and radiation strategies for primary cardiac malignancy.

Declarations

Ethics approval and consent to participate

Not applicable – case reports of a single patient are exempt from ethics committee approval at AdventHealth Orlando.

Consent for publication

Informed consent was obtained from the patient's family for publication of this case report and any accompanying images.

Availability of data and materials

The datasets used and/or analyzed during the current study, including imaging, are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

Funding

Not applicable.

Authors' contributions

APP drafted the manuscript and performed the literature review. TZK and KDA were involved in patient care and critically revised the manuscript. TZK and KDA performed the surgical procedures and reviewed the contents of the manuscript. All authors read and approved the final manuscript.

Acknowledgements

Not applicable.

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Figures

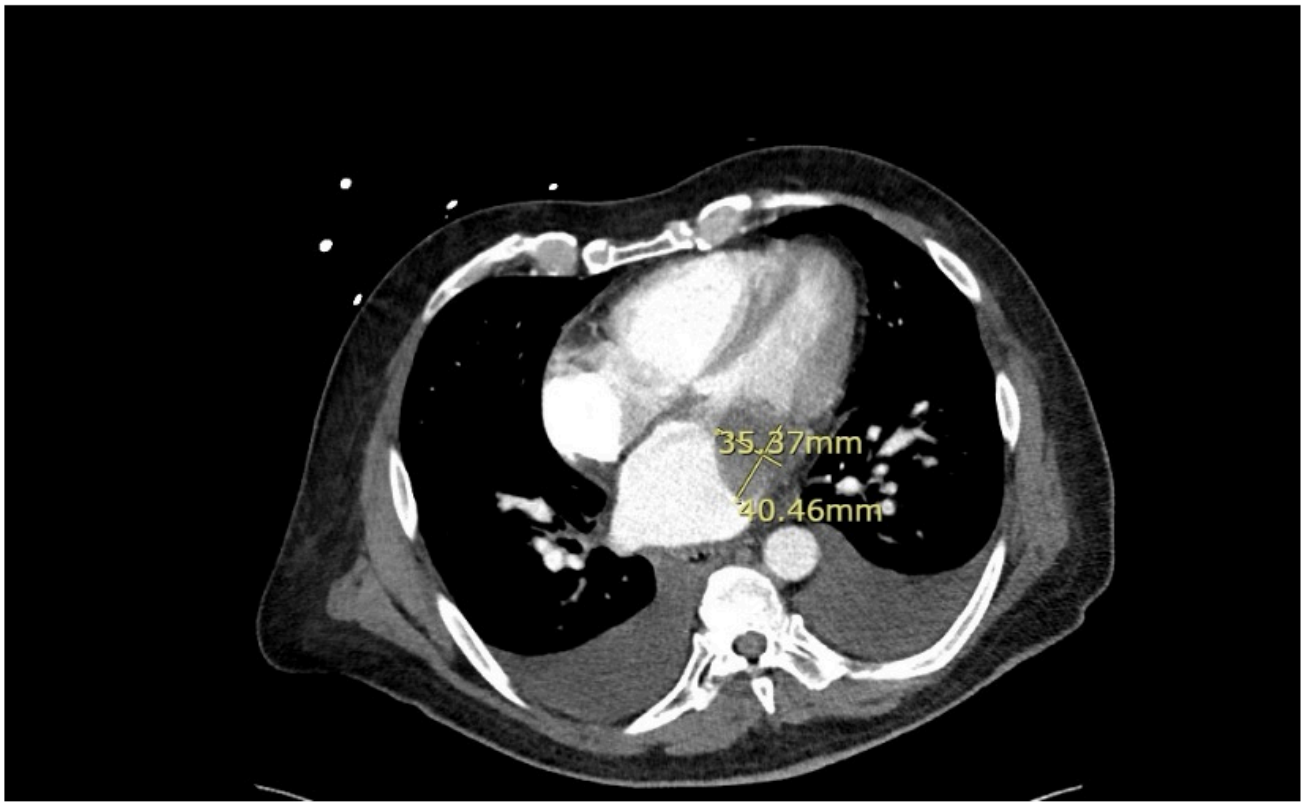


Figure 1

CTA performed two days before surgery demonstrating a 4.1 × 3.5 cm mass along the left atrial wall partially obstructing the left atrial outflow tract.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Video2.mov
- Video1.mov
- Supplemental.docx