An Unusual Cause of Acute Myocardial Infarction Caused by a Large Pulmonary Artery Intimal Sarcoma

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INTRODUCTION

Primary cardiac tumors are rare and have been found in 0.0017%–0.28% of autopsies in the general population.[1] About 25% of primary cardiac tumors are malignant, with cardiac sarcomas accounting for 75% of primary malignant cardiac tumors. Cardiac sarcomas are often locally invasive and fatal if untreated.[1] Several types of primary cardiac sarcomas exist, including right-heart sarcoma, left-heart sarcoma, angiosarcoma, and pulmonary artery sarcoma (PAS).[2,3]

The first case of PAS was reported from an autopsy in 1923.[4] Because of the rarity of PAS, <250 case reports and case series focusing on histopathology and surgical management have been published to date.[4] Herein, we present the case of a 30-year-old man with pulmonary artery (PA) intimal sarcoma, admitted with shortness of breath and subsequently developing an acute anterior wall myocardial infarction.

CASE PRESENTATION

A 30-year-old man presented to the emergency department with progressive exertional dyspnea and pleuritic chest pain. His medical, surgical, social, and family histories were unremarkable; specifically, there were no risk factors for developing premature coronary artery disease or nonischemic cardiomyopathies.

Computed tomography (Figure 1) of the chest revealed a large soft tissue density mass (6.9 × 6.1 cm, 30–60 Hounsfield units), extending from the level of the left atrium and lateral mitral annulus superiorly to the posterolateral aspect of the aortic root and main PA. Transthoracic echocardiography (Figure 2, Videos 1–4) demonstrated a large pericardial effusion with a large extrinsic left atrial mass, adjacent to the main PA and the left sinus of Valsalva. Left ventricular ejection fraction was preserved (67%), without major valvular abnormalities. The inspiratory flow variations across the mitral and tricuspid valves were 58% and 64%, respectively (Figure 3). Cardiac magnetic resonance imaging (Figure 4, Video 5) demonstrated a large mass invading the main and right PAs, resulting in severe intraluminary obliteration. In addition, a large right lower lobe pulmonary infarct was also noted.

The patient was admitted to the cardiac intensive care unit in preparation for surgical resection of the extrinsic cardiac mass. He underwent urgent pericardiocentesis, given pretamponade physiology, to optimize hemodynamics before induction with general anesthesia for surgery. The procedure yielded 260 mL of serosanguinous pericardial fluid, and a pericardial drain was left in situ to gravity for 24 hours.

Four hours following pericardiocentesis, the patient developed an episode of polymorphic ventricular tachycardia, which quickly degenerated into ventricular fibrillation (Figure 5). A cycle of chest compressions (3 min) and unsynchronized cardioversion resulted in successful return of spontaneous circulation. The pericardial drain was removed the next morning, yielding an additional 500 mL of pericardial fluid.

Repeat transthoracic echocardiography at this stage demonstrated resolution of the pericardial effusion. However, left ventricular ejection fraction became significantly depressed (30%–35%), with interval development of severe hypokinesis in the left anterior descending coronary artery (LAD) territory (Video 6). Serum troponin T and creatine kinase-MB were elevated at 1.4 ng/mL (normal range, 0.000–0.029 ng/mL) and 89 ng/mL (normal range, 0.0–2.4 ng/mL), respectively. Emergent left-heart catheterization (Figure 6) revealed an anomalous left circumflex coronary artery arising from the right sinus of Valsalva and a separate LAD ostium from the left coronary sinus, with severe ostial narrowing and angiographic appearance concerning for extrinsic compression. An intra-aortic balloon pump was inserted, and the patient underwent urgent median sternotomy with radical resection of the mass, lymph nodes, PAs, and entire right lung. Intraoperative transesophageal echocardiography demonstrated the proximity of the mass to the aortic root and ostium of the coronary arteries (Figure 7, Videos 7 and 8). Subsequently, a left-sided PA homograft was placed, as well as a reverse saphenous vein graft to the LAD.

The tumor measured 10 × 7.0 × 6.0 cm, with infiltration from the intima of the PA. Pathologic examination (Figures 8 and 9) showed intimal sarcoma with pleomorphism, extending through the PAs and branches into the right lung.

His immediate postoperative course was complicated by circulatory arrest, hemorrhage, and coagulopathy. He remained in cardiogenic shock for 2 days, supported by the intra-aortic balloon pump, inhaled epoprostenol, epinephrine, vasopressors, and an

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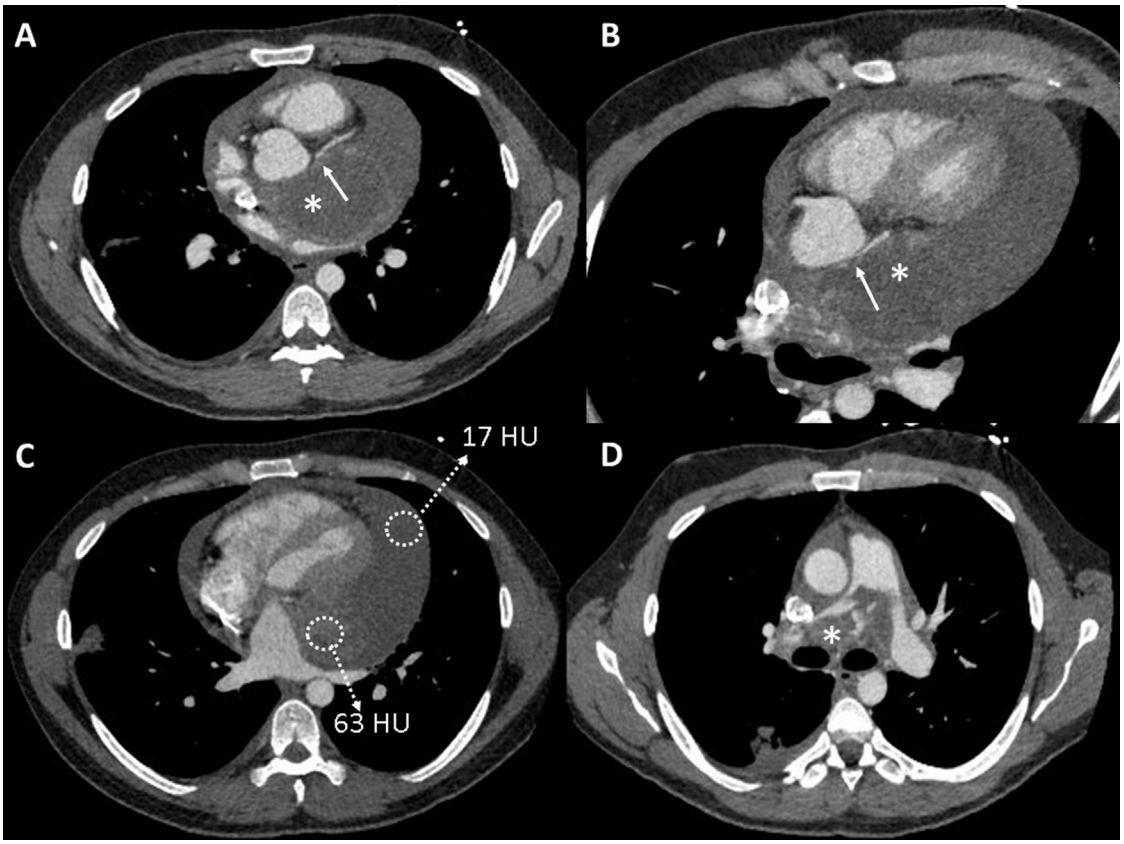

Figure 1 **(A–D)** Cardiac computed tomography. **(A,B)** Multiplanar computed tomographic reconstruction demonstrating severe extrinsic compression of the ostial left anterior descending coronary artery (*arrow*), by the large PA intimal sarcoma (*asterisk*). **(C)** PAS is located lateral to the left atrium and mitral annulus, with a mean attenuation value of 63 Hounsfield units (HU), as opposed to 17 Hounsfield units seen within the pericardial fluid, suggesting high proteinaceous content. **(D)** PAS invading the main PAs, resulting in severe intraluminary obstruction within the right PA (*asterisk*).

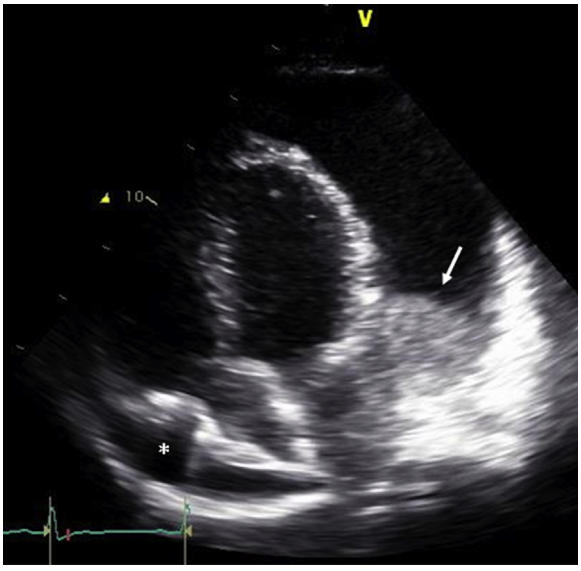

Figure 2 Transthoracic echocardiography, off-axis apical four-chamber view, showing a large circumferential pericardial effusion with pretamponade physiology suggested by systolic right atrial inversion (*asterisk*). In addition, there is a large mass lateral to the left atrium and mitral annulus (*arrow*), consistent with PAS.

open chest. Chest closure and intra-aortic balloon pump removal were successful on postoperative day 2, followed by extubation on postoperative day 4. The patient was discharged on postoperative day 14. At follow-up, the patient remained clinically well. Further treatment with chemotherapy has been planned by the oncology department, to commence as an outpatient.

DISCUSSION

The location of the sarcoma determines not only the clinical presentation but also survival, morbidity, surgical approach, and perioperative mortality.[5] PAS presents with nonspecific signs and symptoms, including chest pain, dyspnea, hemoptysis, cough, constitutional symptoms, and/or right-sided heart failure. Because PAS can be mistaken for PA hypertension, bronchogenic cancer, aneurysm or pseudoaneurysm, or pulmonary embolism that does not respond to anticoagulation, the diagnosis of PAS can be delayed for as long as 3–12 months from the onset of symptoms.[2,3,6,7] A recent case series on pulmonary intimal sarcoma reported dyspnea in all patients (N = 20 [100%]), with chest pain (n = 7 [35%]), constitutional symptoms (n = 5 [25%]), and hemoptysis (n = 3 [15%]) being the other common symptoms.[4] Eighty-five percent of patients (n = 41) reviewed in case reports and case series presented with dyspnea, while 11% presented with cough. Two patients presented

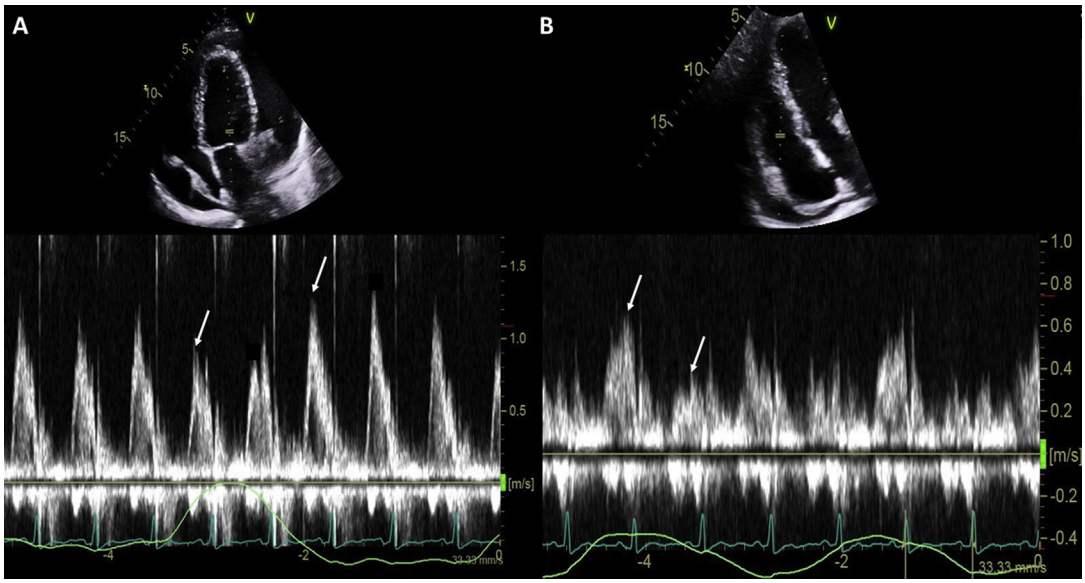

Figure 3 **(A,B)** Transthoracic echocardiography. **(A)** Transmitral pulsed-wave Doppler at the level of the mitral leaflet tips with respirometric tracing, demonstrating significant respiratory variation of >25% when comparing the first beat of inspiration with the first beat of expiration (*arrows*). **(B)** Transtricuspid pulsed-wave Doppler at the level of the tricuspid leaflet tips with respirometric tracing, demonstrating significant respiratory variation of >40% when comparing the first beat of inspiration with the first beat of expiration (*arrows*).

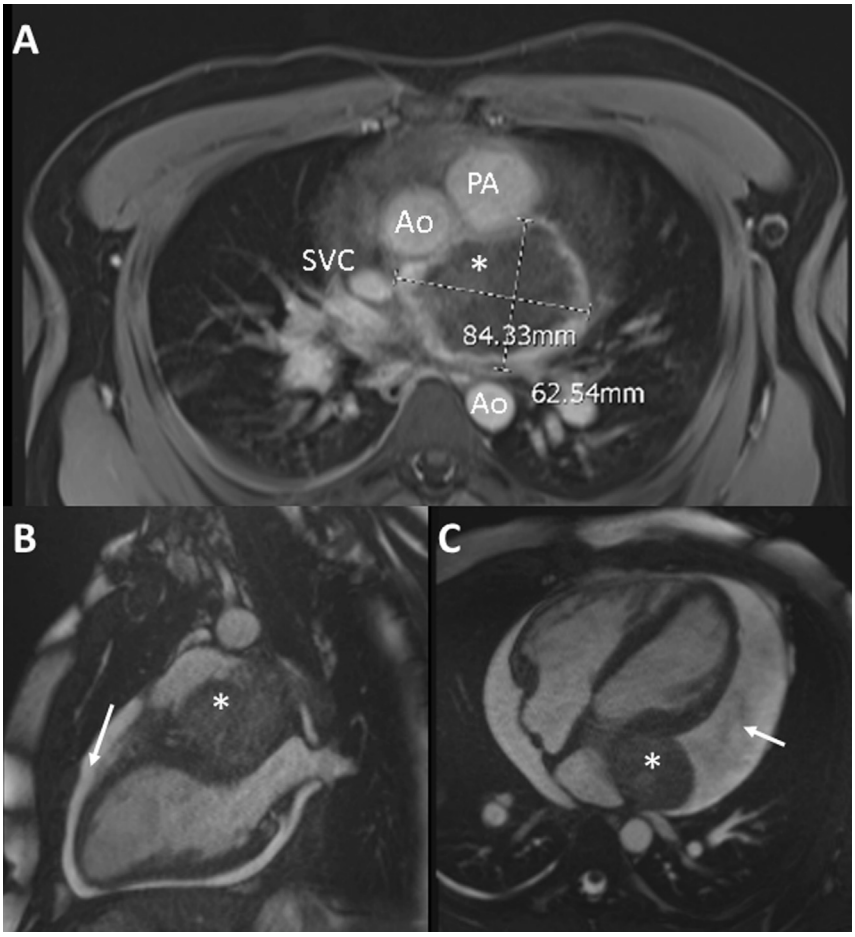

Figure 4 **(A–C)** Cardiac magnetic resonance images demonstrating the size and location of the PA intimal sarcoma (*asterisk*). **(A)** Axial plane through the pulmonary tumor, measuring 8.4 × 6.3 cm in maximal dimensions, and located posterior to the PA and ascending aorta, superior and lateral to the left atrium and ventricle. **(B)** Vertical long-axis view depicting the location of the mass (*asterisk*) in relation to the left ventricle, left atrium, PA, and pericardial effusion (*arrow*). **(C)** Horizontal long-axis view demonstrating a large circumferential pericardial effusion (*arrow*), as well as the tumor mass (*asterisk*), which extends inferiorly and laterally adjacent to the left atrium and mitral annulus (Video 5). *Ao*, Aorta; *SVC*, superior vena cava.

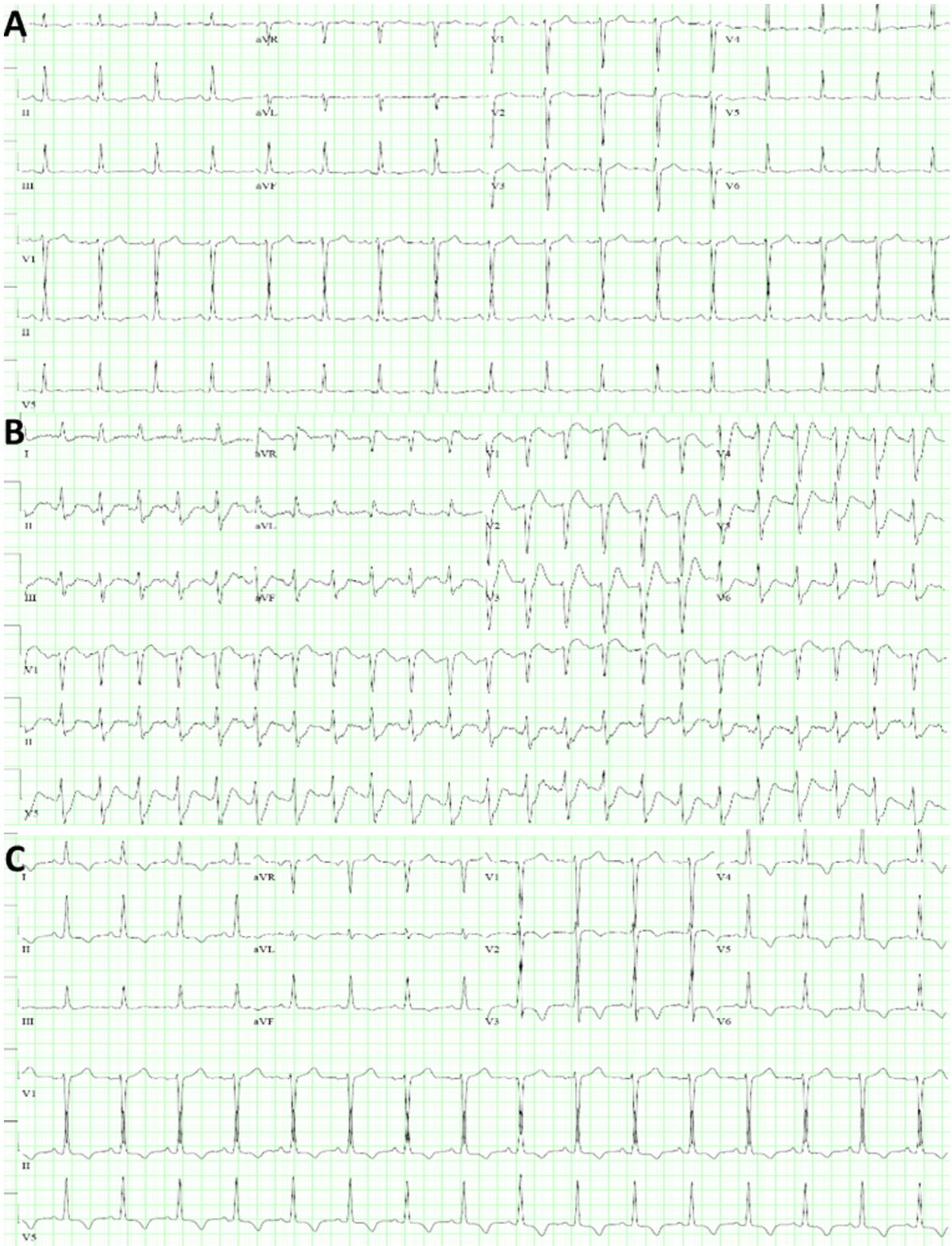

Figure 5 **(A–C)** Electrocardiographic tracings. **(A)** Baseline electrocardiogram demonstrating resting sinus tachycardia with mild electrical alternans in the setting of a large pericardial effusion. **(B)** Electrocardiogram obtained during an episode of resting chest pain, 4 hours after pericardiocentesis. Notice the ST-segment elevations in the anterior leads and lead aVR, with reciprocal inferolateral ST-segment changes. **(C)** Post–myocardial infarction electrocardiogram, demonstrating T-wave inversions in the left anterior descending coronary artery territory with preserved R-wave progression suggestive of viable myocardium. *aVF*, Lead augmented vector foot; *aVL*, lead augmented vector left; *aVR*, lead augmented vector right.

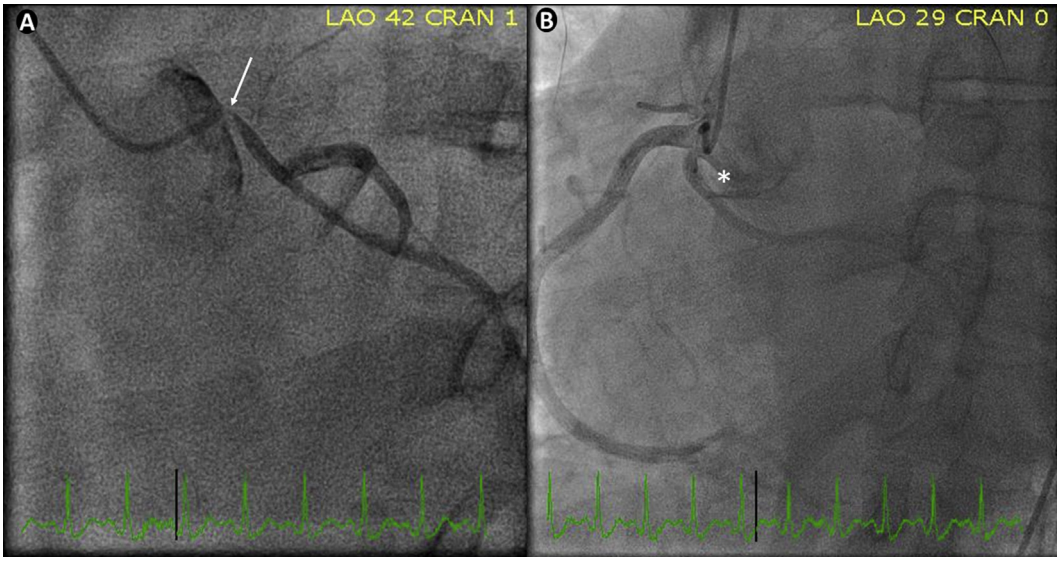

Figure 6 **(A,B)** Coronary angiography demonstrating unusual appearance of the ostial left anterior descending coronary artery segment, suggestive of extrinsic compression of the left anterior descending coronary artery (*arrow*), and an anomalous left circumflex coronary artery (*asterisk*), arising from the right sinus of Valsalva and sharing a common trunk with the right coronary artery. *CRAN*, Cranial; *LAO*, left anterior oblique.

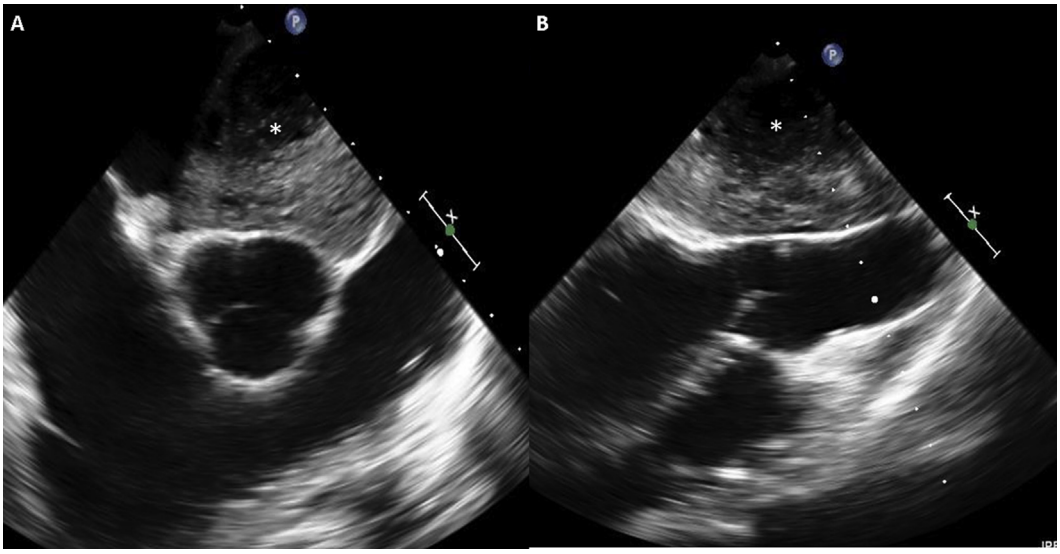

Figure 7 **(A,B)** Intraoperative transesophageal echocardiogram. **(A)** Short-axis view of the aortic valve demonstrating the proximity of the large PA intimal sarcoma (*asterisk*) to the aortic root and ostium of the coronary arteries (Video 7). **(B)** Long-axis view of the aortic valve showing the tumor (*asterisk*) located anterior to the ascending thoracic aorta (Video 8).

with syncope,[8,9] and one case of sudden cardiac death was diagnosed on autopsy.[10]

Transthoracic echocardiography, chest radiography, and computed tomography remain the initial diagnostic tests of choice. Transesophageal echocardiography, following initial transthoracic echocardiography, increases the definition of left-sided masses, while chest, abdominal, and pelvic computed tomography with intravenous contrast allows better assessment of myocardial and pericardial infiltration, as well as extracardiac adjacent and distant metastases.[2,3] Cardiac magnetic resonance provides tissue characterization of a given mass and helps distinguish between matrix and thrombus content, relying on the degree of vascularity and tissue edema.[3] Cardiac magnetic resonance uses specific sequences to differentiate cardiac masses (e.g., PAS) from thrombus, relying on the delayed retention of gadolinium within the extracellular matrix of tumors, which often serves to distinguish between tumor recurrence and postsurgical adjacent mural thrombi.[3,11,12] Cardiac catheterization may be considered to assess tumor burden, particularly in cases in which extrinsic compression of the coronary arteries is suspected.

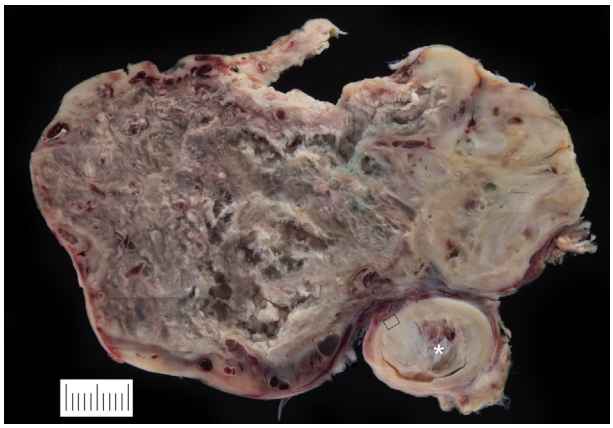

Figure 8 Gross specimen of the tumor mass. The right PA is seen as a discrete transverse section of artery marked by an *asterisk*. The lumen of the artery is almost completely obliterated by tumor. The *small rectangle* marks the area shown in the microscopic field in Figure 9. The intravascular tumor is in continuity with the large extrinsic extracardiac mass. The tumor shows solid areas with fibrous stroma as well as areas of loose connective tissue stroma with spaces created by necrotic tumor. The outer smooth contour of the mass represents containment by a serosal surface. However, there is a ragged area toward the top, where the integrity of the tumor has been compromised. The green dye corresponds to spillover of surgical ink to assess the resection margins.

Tumors from the ventricular myocardium have been reported in the literature to cause arrhythmias and sudden cardiac death because of obstruction to blood flow. In our case, we believe the patient went into ventricular fibrillation arrest soon after pericardiocentesis because of extrinsic compression of the LAD by the large PAS mass, following disruption of the cushioning effect provided by the large pericardial effusion.

Surgical resection involving replacement of the PA and possible pneumonectomy to obtain adequate margins remains the mainstay treatment to prolong survival, though there are no specific guidelines for its management.[4,6,13] PAS is poorly responsive to chemotherapy and radiation, but even though the role of radiation is controversial, the tumor's location above the myocardium possibly makes it amenable to radiation therapy.[6] Doxorubicin and ifosfamide have been reported to be among the most effective chemotherapeutic agents, with survival ranging from 5 to 20 months after treatment, but Penel *et al.* showed no objective stabilization of disease.[14,15] Nevertheless, multimodality treatment has been shown to improve outcomes, with median survival of 24.7 ± 8.5 months compared with 8.0 ± 1.7 months for single-modality therapy.[6]

Given the limited treatment options available, the aggressive nature of the tumor, and delayed diagnosis, prognosis remains poor, with median survival of 36.5 ± 20.2 months after curative surgery compared with 11 ± 3 months for incomplete resection.[6,13,16-20] The common causes of death are predominantly right-sided heart failure from right ventricular outflow obstruction, and distant metastases.[16,17]

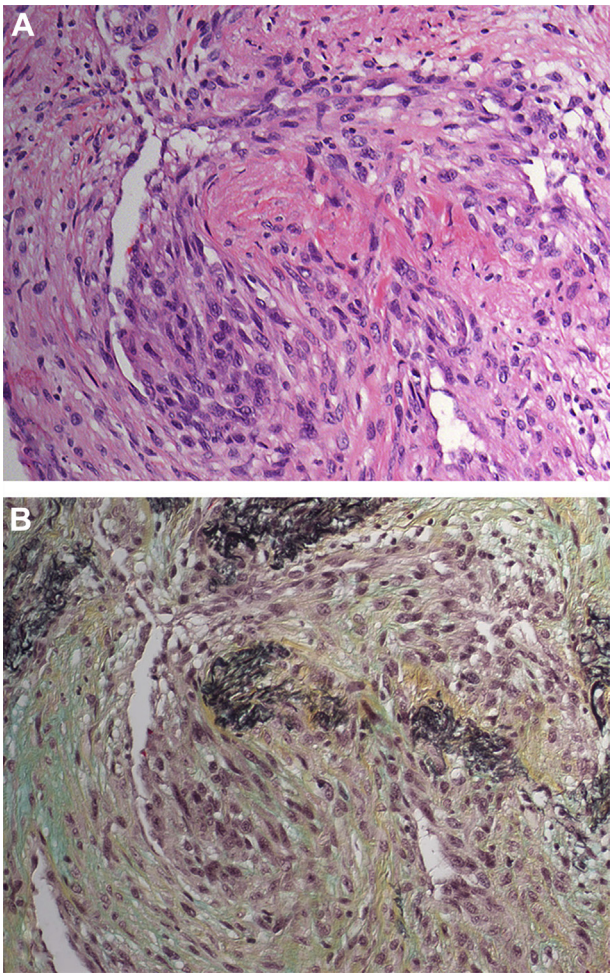

Figure 9 **(A)** Light micrographs of the mass show a pleomorphic, somewhat spindly cellularity with narrow vascular channels corresponding to the nearly obliterated intima on the gross specimen. The *lower image* **(B)** shows a Movat pentachrome stain that further defines the connective tissue of the tumor. It demonstrates fibrous tissue (*yellow*) rich in proteoglycan material (*green*) forming the stroma. This stain also shows destruction of the elastic lamellar units that are usually present in the media layer of the PA. This area of destruction of the media is the area of continuity between the intimal tumor and the large, bulky mediastinal extension (200×, hematoxylin and eosin **[A]** and Movat pentachrome **[B]** stains).

CONCLUSION

The large pericardial effusion likely minimized potential extrinsic compression of the LAD initially, caused by a large PA intimal sarcoma. Subsequent pericardiocentesis and loss of the cushioning effect from the pericardial effusion resulted in additional extrinsic compression of the LAD by the large PA intimal sarcoma, leading to a large anteroapical myocardial infarct. This case highlights the difficult management dilemma created by a large pericardial effusion with pretamponade physiology, caused by a large tumor mass. Clinicians should be mindful of the rare but potentially life-threatening possibility of coronary artery compression by the tumor mass following pericardiocentesis in similar clinical scenarios.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.03.005.

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