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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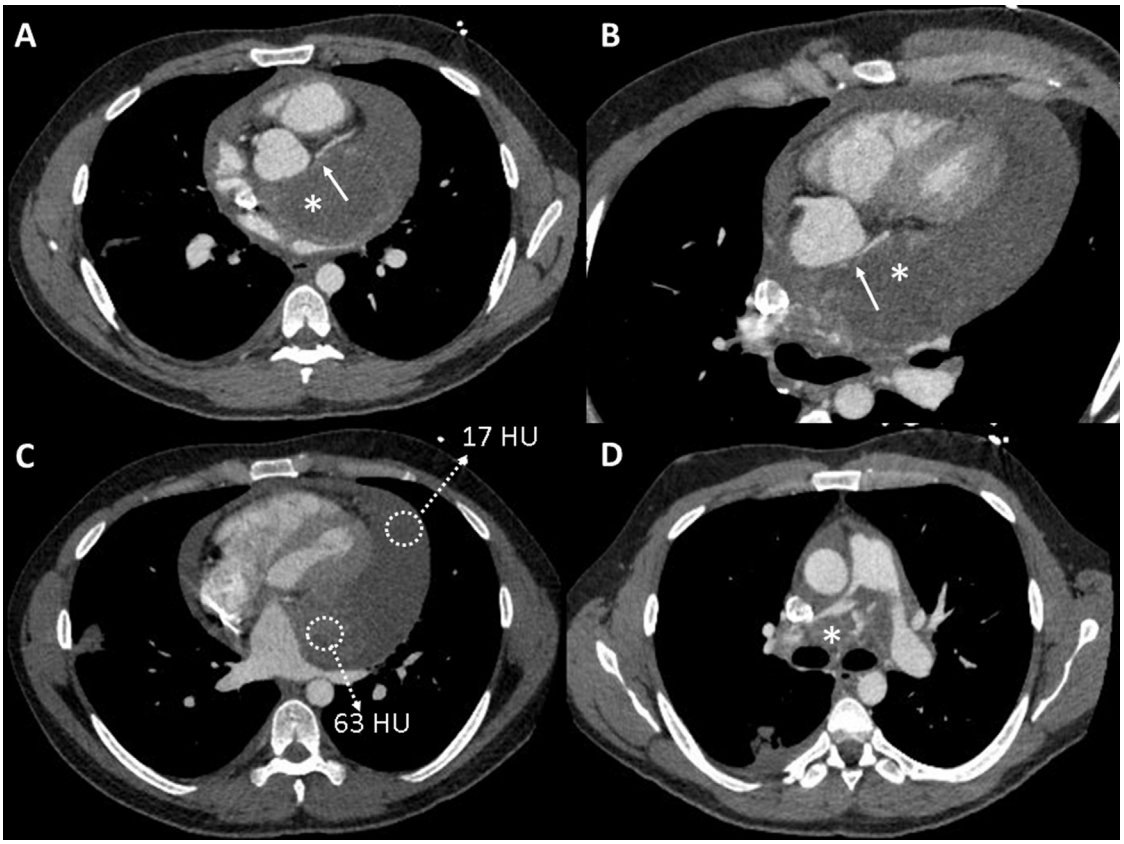

Figure 1 **(A–D)** Cardiac computed tomography. **(A,B)** Multiplanar computed tomographic reconstruction demonstrating severe extrinsic compression of the ostial left anterior descending coronary artery (*arrow*), by the large PA intimal sarcoma (*asterisk*). **(C)** PAS is located lateral to the left 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*).

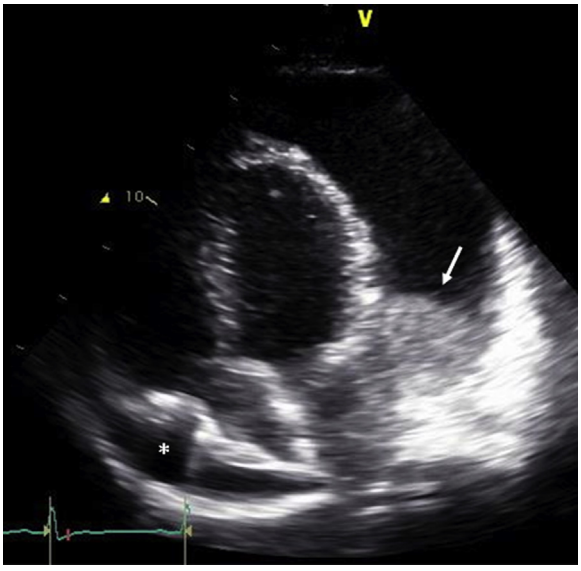

Figure 2 Transthoracic echocardiography, off-axis apical four-chamber view, showing a large circumferential pericardial effusion with pretamponade physiology suggested by systolic right atrial inversion (*asterisk*). In addition, there is a large mass lateral to the left atrium and mitral annulus (*arrow*),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