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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Acute Pulmonary Artery Dissection With an Ongoing Extrinsic Myocardial Infarction

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ABSTRACT

A patient with chronic pulmonary artery hypertension and acute dissection of a main and right pulmonary aneurysm (82 mm) presented with acute myocardial infarction and cardiogenic shock secondary to compression of the left main coronary artery. She required emergency pulmonary artery replacement. She ultimately died due to multiorgan failure and sepsis. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:376–80) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

PRESENTATION

A 65-year-old woman had flu-like symptoms with aching limbs and subfebrile temperatures for a week. Her family physician noticed a progression of symptoms and referred her for extended workup. She went to the emergency room at a community hospital with acute worsening of the symptoms with progressive dyspnea at rest. She further deteriorated, was intubated due to rapidly progressive respiratory failure, and transferred to our institution on inotropic support.

LEARNING OBJECTIVES

- Patients with pulmonary hypertension may develop PAAs.
- PAAs may dissect and rupture.
- PAA and dissection could cause AMI, and eventually, death.

MEDICAL HISTORY

She had a history of resection of a right thigh melanoma 20 years earlier, bronchial hyperactivity, and idiopathic pulmonary arterial hypertension for 25 years. A right-sided heart catheterization was performed at another institution 12 years before, which confirmed severe pulmonary artery hypertension, with a mean pulmonary artery pressure (mPAP) of 55 mm Hg and pulmonary vascular resistance of 900 dynes/s/cm^{-5}. Under inhaled iloprost 10 μg, there was a fall in mPAP to 24 mm Hg and a pulmonary valve resistance of 335 dynes/s/cm^{-5}. She was found to have an aneurysmal main pulmonary artery of 73 × 64 mm on transthoracic echocardiography 6 years ago. She was treated with fenprocumon, spironolactone, amlodipine, bosentan, and tadalafil. No coronary artery disease workup was known to us.

DIFFERENTIAL DIAGNOSIS

At the referring hospital, the suspicion of acute myocardial infarction (AMI) was raised because there

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was ST-T segment elevation on her electrocardiogram and increased troponin T serum levels.

INVESTIGATIONS

On admission, the electrocardiogram showed ST-segment elevation and serum troponin T of 6,475 ng/l, which suggested a ST-segment elevated myocardial infarction. Plain chest x-ray confirmed pulmonary edema. Emergency room transthoracic echocardiography showed a pulmonary artery aneurysm (83.1 mm) as schematically depicted in the **Central Illustration**. The right ventricle was hypertrophic with preserved function. Global left ventricular hypokinesia was confirmed, with an ejection fraction of <25%. A computed tomography scan confirmed acute dissection of the main pulmonary artery extending into the right branch with compression of the left main coronary artery (**Figures 1A and 1B**: main and right pulmonary arteries; **Figures 1C**: 3D reconstruction).

MANAGEMENT

Because the patient was in cardiogenic shock secondary to ST-segment elevated myocardial infarction, after emergency multidisciplinary consultation, salvage surgery was indicated. Intraoperative transesophageal echocardiography confirmed these findings, inclusive of reduced flow in the left main coronary artery. Replacement of the main and right pulmonary arteries with a T-shaped 28-mm prosthetic vascular graft was carried out on cardiopulmonary bypass with beating heart (**Figure 2A**: flow in left main coronary artery; **Figures 2B and 2C**: preoperative; and **Figure 2D**: post-surgery). At completion of the procedure, transesophageal echocardiography confirmed only mild pulmonary insufficiency. She required extracorporeal life support with veno-arterial extracorporeal membrane oxygenation with peripheral cannulation due to left ventricular failure.

She developed multiorgan failure and eventually died from distributive and/or septic shock (positive blood cultures for *Staphylococcus hominis*) on postoperative day 12.

Postmortem examination confirmed chronic fibrosis and sclerosis of the pulmonary arterial tree. There was a myocardial infarction over >30% of the left ventricular mass, extending between the anterior and lateral walls, the interventricular septum, and papillary muscles. There

ABBREVIATIONS AND ACRONYMS

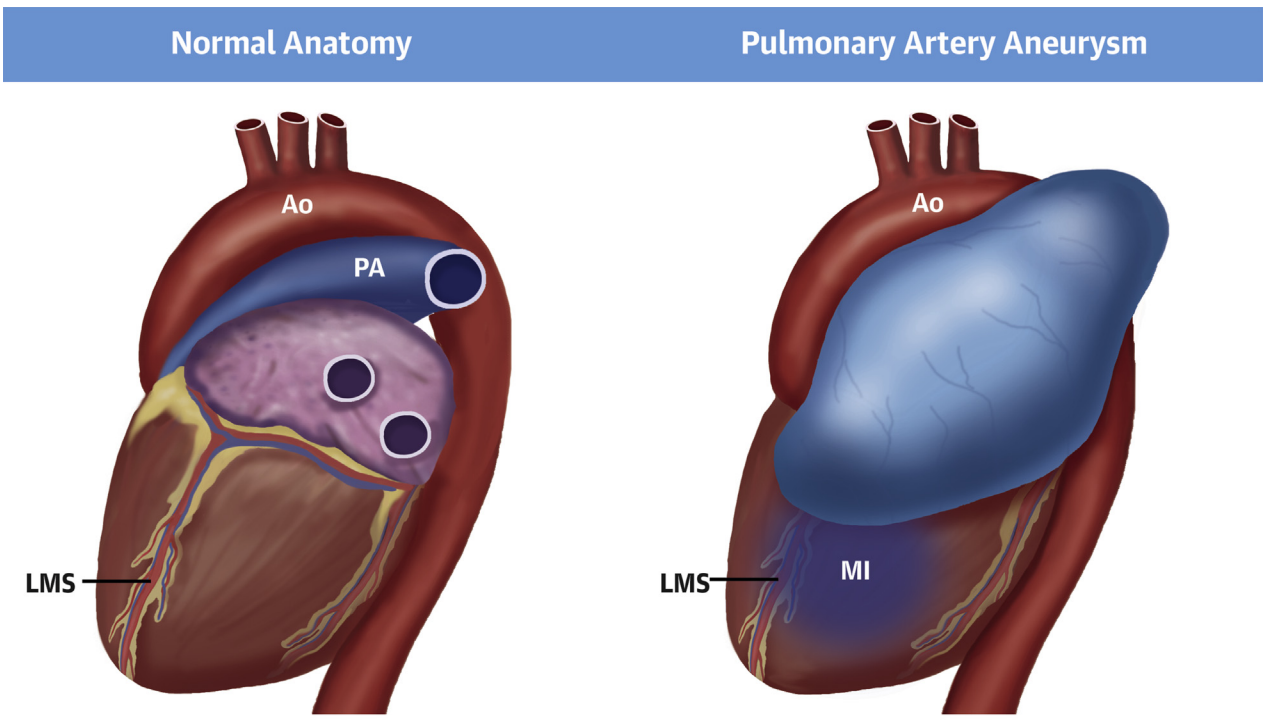
3D = 3-dimensional

AMI = acute myocardial infarction

mPAP = mean pulmonary artery pressure

PAA = pulmonary artery aneurysm

CENTRAL ILLUSTRATION **Schematic Representation of the Location of the Pulmonary Artery Aneurysm**



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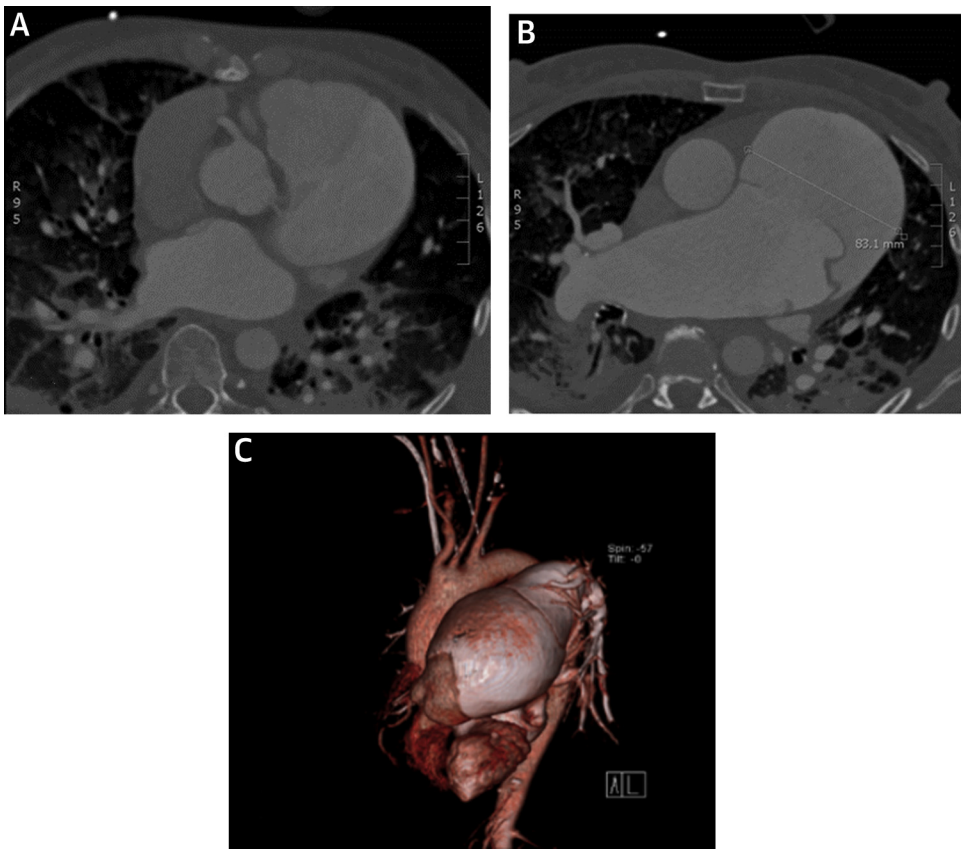
Ao = aorta; LMS = left ventricular mass; MI = myocardial infarction; PA = pulmonary artery.

FIGURE 1 Preoperative Imaging of the Dissected Pulmonary Artery



(A) Enlarged pulmonary artery, dissection membrane on the medial side, pulling sideways toward bifurcation in the cranial aspect. Left coronary bifurcation was involved. **(B)** Main pulmonary artery aneurysm and dissection. Main trunk maximal diameter was 83.1 mm. **(C)** 3-dimensional reconstruction.

was occlusion of the left bronchial tree with fresh clot.

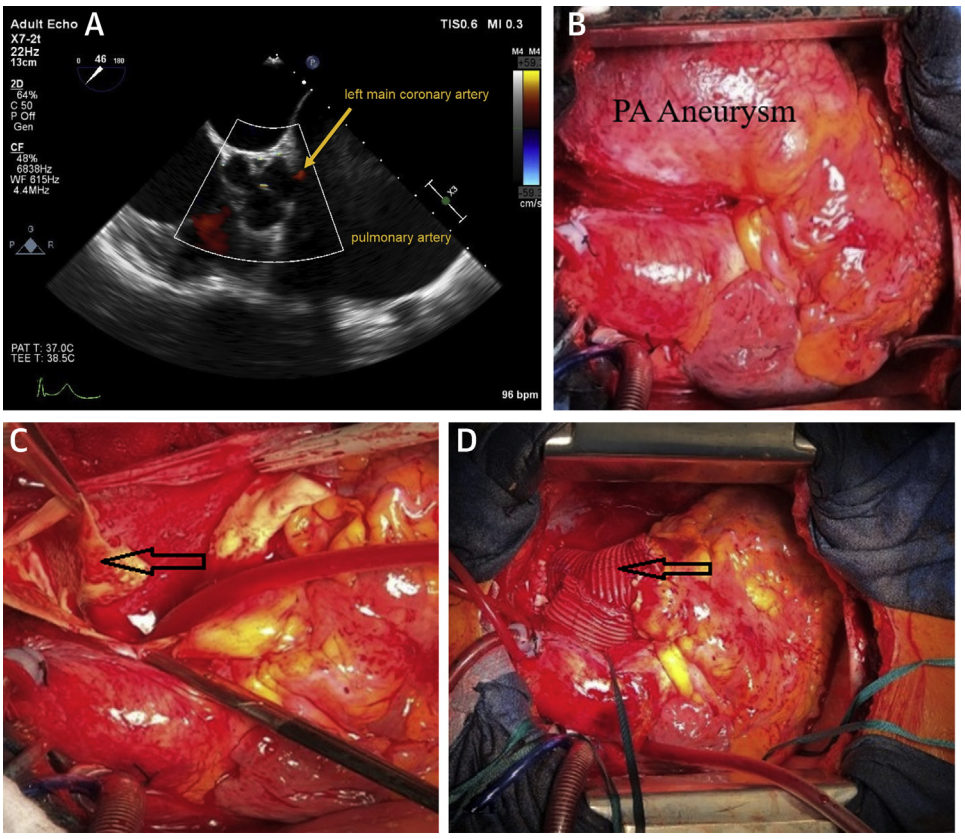
The case was considered cardiac insufficiency secondary to diffuse myocardial infarction and cardiogenic shock in combination with acute respiratory insufficiency with occlusion of the left bronchial tree due to clot.

DISCUSSION

Pulmonary artery aneurysm (PAA) is an uncommon entity in adults and is related to congenital cardiac and vascular anomalies, as well as acquired diseases like pulmonary artery hypertension (1,2). PAA has been described in relation to cystic medial necrosis (3). In congenital heart disease, PAA has also been associated with left-to-right shunts as in patent ductus arteriosus ventricular and atrial septal defects, and as a long-term post-operative sequelae of congenital heart disease repair. Idiopathic PAAs also exist (4,5). Due to the availability of diagnostic imaging methods, there has been an increase in reports of such cases. Morphologically, PAA is described as a focal dilatation involving all 3 layers of the vessel wall. The main causes of mortality with PAA are dissection and rupture (6-9). On computed tomography scans, the upper normal diameter limit of the main pulmonary artery is 29 mm and that of the right interlobar artery is 17 mm. PAAs that exceed these limits are considered enlarged, and one that exceeds 4 cm is considered aneurysmal (2,6,10).

Because this is a rare condition, it is difficult to confirm a threshold for operability. Per their

FIGURE 2 The Sequence of Surgical Treatment



(A) Intraoperative transesophageal echocardiography view of left main coronary artery flow. **(B)** intraoperative view of main pulmonary artery (PA) aneurysm. **(C)** Membrane dissection **(arrow)**. **(D)** Surgical reconstruction: main and right pulmonary vascular grafts.

literature review, the authors estimated that a little >100 cases have been described, with approximately 25% diagnosed at post-mortem examination. The association with AMI might be even rarer. Some investigators advocate an operation as soon as PAA has been diagnosed to prevent rupture, whereas others recommend surgery for all PAAs with a diameter ≥60 mm (11–13). In this case, salvage operation was deemed the only treatment option for a patient in cardiogenic shock due to AMI.

CONCLUSIONS

AMI caused by the compression of the left coronary artery due to acute pulmonary artery dissection is uncommon. PAAs may also be dissected.

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KEY WORDS **acute myocardial infarction, pulmonary artery aneurysm, pulmonary artery dissection**