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Case Report

Pulmonary artery dissection in long standing idiopathic pulmonary arterial hypertension: A case report [☆]

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ABSTRACT

Pulmonary arterial dissection (PAD) is a rare and often lethal complication of chronic pulmonary arterial hypertension (PAH), which may occur in patients with idiopathic pulmonary arterial hypertension (IPAH) and potentially in those with connective tissue disorders. While rare, sudden death often occurs secondary to acute cardiac tamponade, as the pulmonary artery dissects into the pericardium; this diagnosis is often made postmortem. Nevertheless, with the proliferation of multidetector computed tomography (MDCT) as a diagnostic test, patients may be identified very early after symptom onset, prompting rapid intervention with decreased morbidity and mortality. We report a case of IPAH complicated by pulmonary artery aneurysm (PAA) and PAD, diagnosed by CT pulmonary angiogram (CTPA), and treated with bilateral lung transplantation, pulmonic valve replacement, and re-anastomosis of the donor main PA to a pulmonary valve conduit.

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Case report

A 35-year-old woman with a past medical history of idiopathic pulmonary arterial hypertension and pulmonary artery aneurysm (Fig. 1) presented to the emergency department af-

ter awakening with severe chest pain. Additional pertinent medical history included heart failure with preserved left ventricular ejection fraction, hypercoagulability secondary to Factor V Leiden, systemic lupus erythematosus, hepatomegaly, and chronic respiratory failure. The acute chest pain was described as left sided, worse when lying on her left side, and

[☆] Competing interests: Dr. Ganapathi has served as a consultant for Abbvie Pharmaceuticals in the past year. This activity is not relevant to the current endeavor. Otherwise, None.

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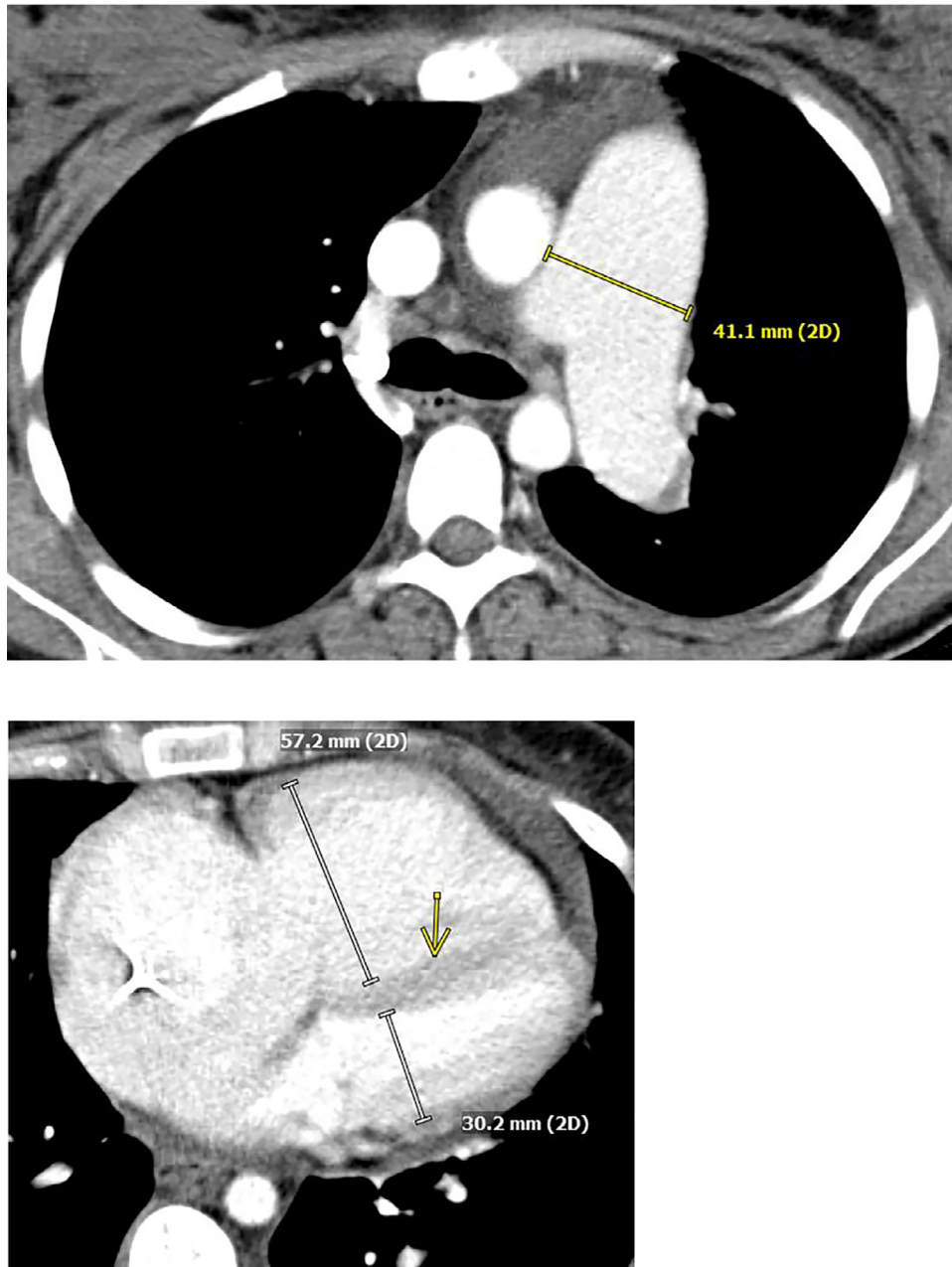


Fig. 1 – CT pulmonary angiogram 2007: (A) Axial CT image at the level of the carina reveals an enlarged main pulmonary artery measuring up to 4.1 cm, consistent with a pulmonary artery aneurysm. **(B)** Axial CT image at the level of the right atrium demonstrating enlargement of the right atrium and ventricle, with subtle leftward deviation of the interventricular septum, findings suggesting right heart strain. Also demonstrated is a small pericardial effusion.

pleuritic in nature. Based on the clinical history and presenting symptoms, the differential considerations included pulmonary embolism, aortic dissection, myocardial infarction, pericardial effusion, or acute injury of the main pulmonary artery (including rupture/dissection).

On arrival the patient was found to have an elevated D-dimer. She was tachycardic and tachypneic on examination, but had regular rate and rhythm without any murmurs, rubs, or gallops on cardiac auscultation. The breath sounds were normal on auscultation bilaterally. She had no lower extremity edema or other signs of volume overload. A chest radio-

graph was unchanged from 5 months prior, and demonstrated gross enlargement of the cardiomeastinal silhouette, and near opacification of the left hemithorax (Fig. 2). A follow-up CT pulmonary angiogram revealed a markedly dilated main pulmonary artery measuring up to 11.1 cm, with an intimal flap/dissecting septum (Fig. 3). Also present was an associated high attenuation (HU 35–45) hemorrhagic pericardial effusion. There were no signs of pulmonary embolism. A study from 5 months prior revealed that the main PA had measured 9.5 cm (Not shown), without a dissecting septum. A follow up echocardiogram revealed right heart strain with right atrial

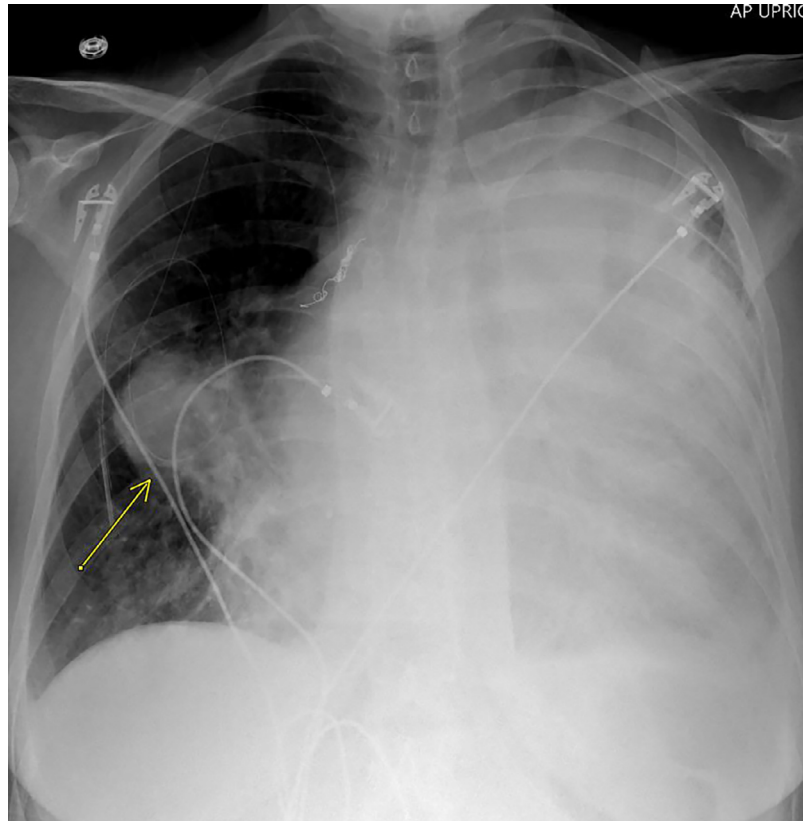


Fig. 2 – PA chest radiograph 2021. Near complete opacification of the left hemithorax, secondary to a grossly enlarged main pulmonary artery and its continuation into the left pulmonary artery, and associated enlargement of the cardiomeastinal silhouette. Enlargement of the right pulmonary artery and the right interlobar pulmonary artery (yellow arrow).

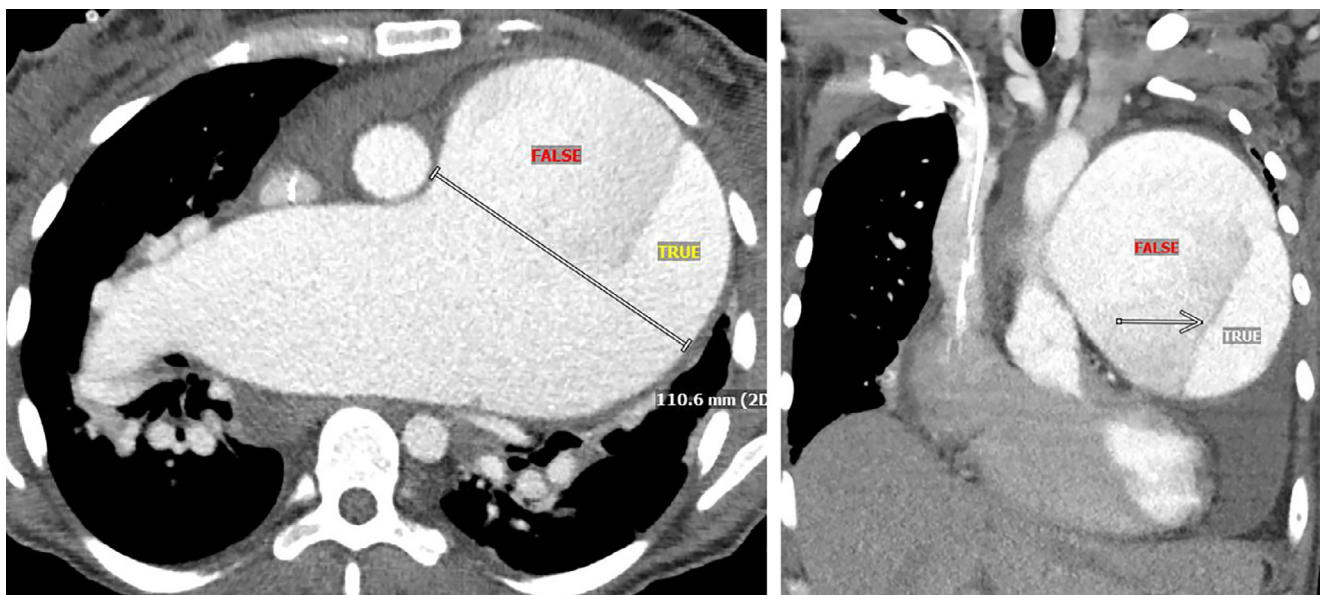


Fig. 3 – CT pulmonary angiogram 2021: (A) Axial CT image at the level of the right pulmonary artery demonstrates a marked pulmonary artery aneurysm measuring up to 11.1 cm, with a dissection septum separating the false and true lumen. Also noted is enlargement of the right pulmonary artery and an associated moderate sized pericardial effusion. (B) Coronal image again demonstrates the pulmonary artery aneurysm and dissection, and also a moderate sized pericardial effusion. The enlarged cardiomeastinal silhouette [Figure 2](#), is likely related to the pericardial effusion and pulmonary artery aneurysm.



Fig. 4 – Gross image of the internal lumen of the pulmonary artery aneurysm with an acute and chronic dissection. The forcep at the bottom of the screen is pointing to the thrombus within the false lumen (a).

and ventricular chamber size enlargement, and a marked pulmonary artery aneurysm measuring 11.4 cm in diameter, with a linear echogenic density consistent with the dissecting septum. Troponin and brain natriuretic peptides were within normal limits. The patient was hospitalized for marked pulmonary artery aneurysm and dissection management.

Multidisciplinary evaluation by Cardiology, Pulmonology, and Cardiothoracic Surgery was initiated. The patient was already listed for bilateral lung transplantation for chronic IPAH and PAA, prior to the acute presentation. The consensus decision was taken to proceed with bilateral lung transplantation. Prior to surgery, peripheral VA-ECMO was implemented to control pulmonary arterial hypertension and decompress the right heart. Following this intervention, the patient underwent bilateral lung transplantation with excision of the aneurysmal and dissected pulmonary arteries and the native pulmonary valve. The gross specimen revealed a dilated pulmonary artery measuring $11.8 \times 10.2 \times 5$ cm. The intimal surface was partially disrupted, the dissection measured 7.2×5.2 cm, with partial thrombus formation in the false lumen (Fig. 4). The patient had the pulmonary valve replaced with a 25 mm Medtronic (Minneapolis, MN) Hancock Bioprosthetic Valve Conduit with anastomosis of the donor pulmonary artery to the graft conduit. Post-transplant chest radiograph is displayed in Figure 5. Following transplantation, a pathological examination of native lung specimens confirmed the diagnosis of PAH. The right and left explants revealed pulmonary arteriopathy with concentric laminar medial hypertrophy and plexiform lesions, in keeping with long standing PAH.

The patient was then discharged to a skilled nursing facility and completed Pulmonary Rehabilitation sessions at a local facility. The patient is currently doing very well, off oxygen and is back at work.

Discussion

Pulmonary hypertension (PH) represents a complex pathophysiological condition, with several clinical entities resulting

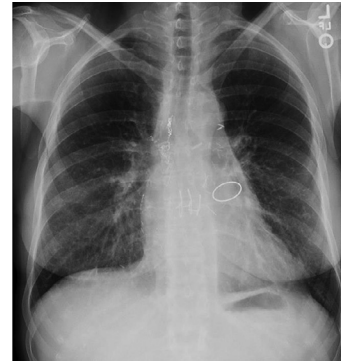


Fig. 5 – PA chest radiograph postbilateral lung transplantation and pulmonic valve replacement. Evidence of clamshell sternotomy, bilateral lung transplantation, embolization of the right internal mammary artery, and pulmonic valve replacement. The cardiomeastinal silhouette is normal in size.

in increased pressure in the pulmonary circulation, and progressive impairment of the cardiopulmonary function which typically results in right ventricular failure [1]. Regardless of the underlying mechanism, PH is defined by a mean pulmonary arterial pressure greater than 25 mmHg. On CT imaging PH presents as an enlarged/ectatic main pulmonary artery (main PA diameter > 3 cm), with or without signs of right heart strain or dysfunction [2–4]. Pulmonary artery aneurysm (PAA) is considered when the main pulmonary artery diameter is greater than 4 cm. PAA formation and progression is associated with increased vessel wall stress and the potential of rupture.

Pulmonary arterial hypertension (PAH) is a chronic lung disease characterized by progressive and pathologic remodeling of the resistance in the pulmonary arteries [1,3–6]. This group of disorders is described under the WHO group 1 classification of PH [7]. Idiopathic pulmonary arterial hypertension (IPAH) is diagnosed in patients with no identifiable cause. This condition accounts for 39% of PAH cases (Bazan), predominantly affects young women, and has a poor prognosis. IPAH in patients refractory to medical therapy, is a clinical indication for lung transplantation. Development of a markedly enlarged (marked) PAA is a complication of long standing IPAH [7].

Pulmonary arterial dissection (PAD) is a rare and often lethal complication of chronic PAH. Patients may present with chest pain, worsening dyspnea, cyanosis, hemoptysis, cardiogenic shock, or sudden death. The main pulmonary artery is the sight of dissection in approximately 80% of cases [8,9]. The remainder of cases are limited to the right and left pulmonary artery, and the interlobular and segmental pulmonary artery branches. Additional associations with PAD include focal PAA, pulmonary artery infection/inflammation, cardiac amyloidosis, trauma, and connective tissue disease. In contrast to aortic dissection, the false lumen in pulmonary artery dissection tends to rupture, rather than extend distally or develop a re-entry tear [8], causing exsanguination and hemodynamic instability of the patient often leading to death. Sudden death

may occur secondary to acute cardiac tamponade, as the pulmonary artery dissects into the pericardium. In addition to the pericardium, other sites of extension include into the lung parenchyma, mediastinum, and pleural space, also resulting in sudden death. An increased risk for PAD and unexpected death has been reported for pulmonary artery diameters measuring greater than 4.8 cm [10].

Most cases of PAD have been diagnosed at autopsy. There are increasing reports of diagnosis of PAD with noninvasive imaging modalities including transthoracic echocardiography (TEE), contrast enhanced MDCT, and magnetic resonance imaging (MRI). Echocardiography is a noninvasive screening tool for PH. However there is varying sensitivity and specificity of this modality for detecting PAD, as it is operator dependent and may be limited by other factors including patient body habitus. Slow blood flow may be present in cases of PAH, obscuring evaluation of a dissecting septum in a suspected case for PAD, on MRI examinations. In addition, MRI examinations are more time consuming, lack the spatial resolution of MDCT, and are generally more expensive than MDCT.

Contrast enhanced MDCT, specifically ECG-gated CTPA is a rapid and robust non-invasive diagnostic tool essential to diagnosis and treatment planning in the management of pulmonary artery dissection. The advantages of CTPA include its widespread availability, the speed of image acquisition, its high spatial resolution, and the ability to perform postprocessing techniques. ECG gated CTPA will decrease motion artifact which mildly limits the visualization of the right ventricular outflow tract. Multiplanar reformation, maximum intensity projection, and 3-D imaging may also optimize the evaluation of marked pulmonary artery aneurysm and pulmonary artery dissection. In addition, right heart strain and other imaging complications including the presence of a pericardial effusion, lung parenchymal hemorrhage, or pulmonary embolism may be demonstrated.

Treatment of patients with PAH, specifically IPAH includes supportive therapy, drugs (anticoagulation, diuretics, Prostaglandins, Sildenafil, endothelin receptor agonists), and lung transplantation in patients with progressive/refractory disease. Lung transplantation is indicated in patients with long-standing pulmonary arterial hypertension. In this case complicated by the presence of a marked PAA and PAD, additional pulmonary valve replacement and placement of the proximal conduit was performed. Alternatively combined lung and heart transplantation has also been performed to treat long standing IPAH complicated by a marked PAA [6]

This case report described IPAH in a young (39 years) female, complicated by development of a marked PAA, which was further complicated by development of an acute life-threatening PAD. This case demonstrated diagnosis by CTPA, which identified the dissecting septum, the extent of dissection, the size of the main pulmonary artery, and a moderate sized pericardial effusion. After multidisciplinary evaluation, management with bilateral lung transplantation, pulmonic valve replacement with an associated conduit, and anastomosis

of the donor main pulmonary artery to the conduit, was successfully performed.

In conclusion, PAD is a rare and often fatal condition affecting patients with chronic PAH and a marked PAA, and should be suspected in the patient's presentation with new chest pain, back pain, worsening dyspnea, cyanosis, and hemodynamic compromise. CTPA is an excellent widespread, rapid, noninvasive imaging modality used to identify PAD and potential associated complications. Rapid diagnosis and surgical intervention decreases morbidity and mortality associated with this rare condition.

Informed consent

The authors obtained consent from the patient for submission of the case findings for publication.

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