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CASE REPORT

Pulmonary Artery Dissection (PAD): A Very Unusual Cause of Chest Pain

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KEYWORDS: chest pain, pulmonary artery dissection, pulmonary hypertension.

A 51-year-old African American woman with medical history of essential hypertension and chronic obstructive pulmonary disease (COPD) presented to the hospital with chest pain and shortness of breath. The chest pain was retrosternal and radiated to the back. It lasted for about an hour and resolved without any intervention. After some time, she again felt discomfort in the chest, which was a constant and dull ache.

She had similar episodes of chest pain 1 week prior, although less severe in intensity, for which she went to an outside hospital before coming to our hospital. Acute coronary syndrome was ruled out with serial cardiac enzymes measurements. An exercise stress test was also performed at that time, which failed to show any stress-induced ischemia.

Her medications included lisinopril for hypertension and aspirin, which had been started 1 week prior to admission. She gave a 10-pack-year history of smoking tobacco. Family history was significant for hypertension in her father and coronary artery disease in her mother at the age of 58 years. A review of systems was negative for fever, cough, orthopnea, wheezing, palpitations, nausea, vomiting, recent surgery, or any significant trauma.

Assessment

The patient's physical examination was only remarkable for a blood pressure (BP) of 181/100 mm Hg. She did not have Marfanoid features, hyperflexible joints, or easy bruisability. Laboratory tests, including complete blood count, comprehensive metabolic panel, and cardiac enzymes, were within normal limits. A contrast-enhanced computed tomography (CT) of the chest showed a linear hypodense area in the left lateral aspect of the main pulmonary trunk, which suggested an intimal dissection of the main pulmonary artery. Magnetic resonance angiography/imaging (MRA/MRI) confirmed dissection of the main pulmonary artery extending into the proximal left pulmonary artery and associated with a 12×8 mm^2 aneurysm (Figures 1 and 2). The entry site of dissection was located in the main pulmonary artery just after its origin and the exit site was located in the left pulmonary artery 5 mm distal to the bifurcation of main pulmonary artery. The pulmonary artery diameter at the dissection was 27 mm.

Diagnosis

To investigate possible etiologies, a transthoracic echocardiogram with Doppler was done to look for pulmonary hypertension. The echocardiogram showed normal pulmonary artery pressure with normal right ventricular systolic pressure. There was no evidence of pericardial effusion or structural cardiac abnormality on echocardiogram. Further investigations including work up for connective tissue diseases and infectious etiologies (Table 1) were normal.

According to Shilkin et al.,[1] Helmbrecht first reported pulmonary artery dissection (PAD) in 1842. PAD is very rare and is usually diagnosed at autopsy. There are 71 other cases of PAD reported in the English literature, of which 16 cases are in living patients.[2–17] Unlike aortic dissection, which is fairly common, the reentry circuit for PAD is formed by the rupture of the free wall of the pulmonary artery leading to hemopericardium, cardiac tamponade, and sudden death.[2,8,9] There is wide variation in age of incidence, ranging from 26 to 85 years of age, with a slightly higher prevalence in females (male-to-female ratio 1:1.2).[1,2] The main pulmonary artery is usually involved, with or without involvement of its branches. Isolated left and right pulmonary artery involvement is seen in 6% and 4% of cases, respectively.[2]

Pulmonary hypertension, either primary or secondary (collagen vascular diseases, COPD, congenital heart diseases, etc.), is the most common underlying etiology. Other less common, but well-described etiologies include: Marfan's syndrome, instrumentation of pulmonary artery, tuberculosis, syphilis, pregnancy, idiopathic cystic medial necrosis, and amyloidosis.[2,8]

As noted earlier, underlying pulmonary hypertension is usually a major risk factor for PAD. More than 75% of the patients have underlying acute or chronic pulmonary hypertension.[2] Our patient had COPD without pulmonary hypertension. Despite extensive investigation, no underlying cause of her pulmonary dissection was identified. The

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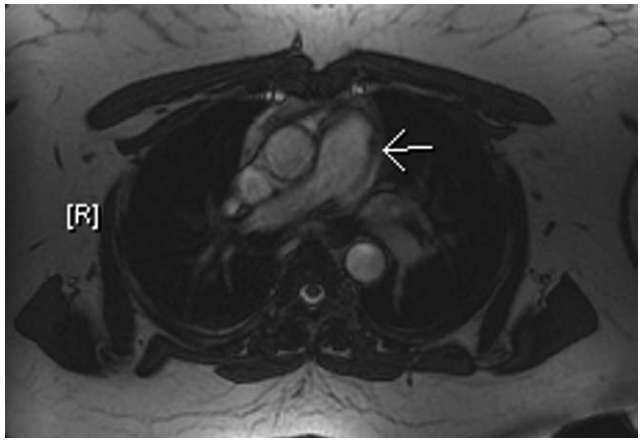

FIGURE 1. Cardiac MRI showing intimal flap involving the left lateral aspect of the main pulmonary artery and partially extending across the opening of the left pulmonary artery. Abbreviation: MRI, magnetic resonance imaging.

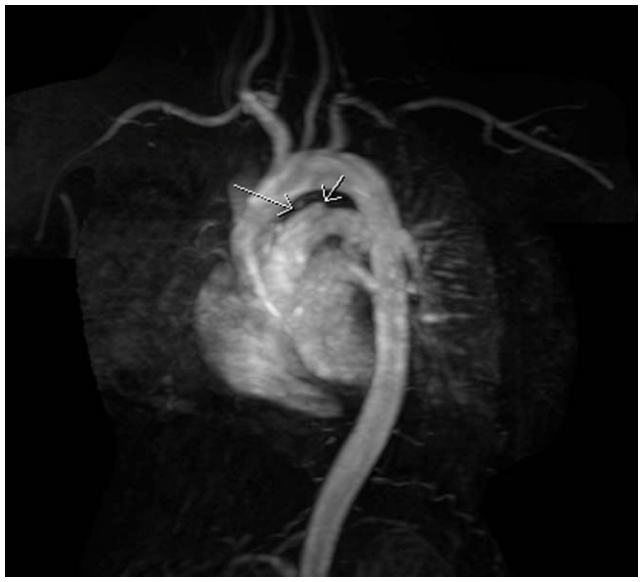

FIGURE 2. Cardiac MRA showing intimal flap involving the left lateral aspect of the main pulmonary and associated 12 mm × 8 mm aneurysm involving superior aspect of the main pulmonary artery and proximal left pulmonary artery. Abbreviation: MRA, magnetic resonance angiography.

differential diagnosis includes cryptogenic cystic medionecrosis; however, because the patient refused surgery the diagnosis remains unknown. As in our case, idiopathic PAD is extremely rare, and only 4 other cases have been described in the literature.[2] Underlying etiologies should always be ruled out to identify correctable causes such as congenital abnormalities of the heart leading to pulmonary hypertension.

Chest pain is a very common presenting complaint in the emergency department. Although rare, PAD should be suspected in a patient with retrosternal chest pain when other common causes of chest pain are excluded. Some of the more suggestive findings are the presence of a new diastolic murmur, a wide mediastinum on chest x-ray, and CT scan of chest showing an intimal flap.[2,8] CT scan of the chest is an acceptable imaging modality to diagnose PAD.[18] According to Neimatallah et al.,[18] there are only 5 cases in the literature reported with detailed CT scans demonstrating PAD. If the diagnosis remains uncertain, it should be confirmed by MRI/MRA.[16] Transthoracic echocardiography can be used for diagnosis and follow-up of conservatively managed patients with PAD.[3,8,19] The echocardiographic findings suggestive of PAD include direct or indirect evidence of pulmonary artery hypertension, with a large main pulmonary artery and an intimal flap across the pulmonary trunk.

TABLE 1. Collagen Vascular Disease and Infectious Disease Workup

Variable	Reference Range	Patient's Result
ANA	Negative	Negative
C3 complement level	88–201	175
C4 complement level	16–47	49
RF	<20	<20
Anti-centromere Ab	Negative	Negative
Anti-Scl 70 Ab	Negative	Negative
Anti-smooth muscle Ab	Negative	Negative
Anti-mitochondrial Ab	Negative	Negative
Anti-parietal cell Ab	Negative	Negative
TB skin test		5 mm
RPR	Negative	Negative

Abbreviations: Ab, antibody; ANA, antinuclear Ab; RF, rheumatoid factor; RPR, rapid plasma reagin; Scl, scleroderma; TB, tuberculosis.

Management

No consensus strategy is available for the management of PAD because of the rarity of this condition. In general, operative repair is the treatment of choice for PAD.[2,8,9,11] There are 16 cases diagnosed in living patients, out of which 6 were managed medically, 8 were managed surgically, and management was not discussed in 2 of the case reports (Table 2). In these case reports, medically managed patients were treated with oxygen, vasodilators (nitrates, angiotensin-converting enzyme [ACE] inhibitors, dihydropyridine calcium channel blockers, sildenafil), diuretics and beta-blockers. These patients did well on follow-up, ranging from 3 weeks to 4 years, except for 1 who died on day 4 in the intensive care unit (ICU).

Conservative management may be tried in patients who are hemodynamically stable and do not have pericardial effusion.[2,9] The aim of conservative management is to decrease right ventricular preload and afterload. Preload reduction can be dangerous in patients with PAD and should be done in the intensive care setting as this can lead to profound hypotension. Nitrates for preload reduction

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TABLE 2. Case Report Descriptions of PAD in Living Patients, With Underlying Etiology, Management, and Outcome

Case Report	Etiology of PAD	Management	Outcome
Janus et al.[3]	Balloon valvuloplasty for pulmonary stenosis	Medical (beta blocker)	Stable during 4 years of follow-up
Khattar et al.[8]	Secondary PH from COPD	Medical (diuretics, ACE inhibitor)	Stable during 1 year of follow-up
Lobato et al.[9]	Aortic valve replacement	Medical (vasodilators, diuretics)	Stable during 3 weeks of follow-up
Smalcelj et al.[10]	Primary PH	Medical (Sildenafil)	Stable during 8 months of follow-up
Song and Kolecki[11]	Secondary PH from VSD, Eisenmenger's syndrome	Medical (Nitroprusside)	Patient died on day 4 of admission
Steurer et al.[15]	Primary PH	Medical (ACE inhibitor, CCB)	Stable during 1 year of follow-up
Wuyts et al.[4]	Secondary PH from VSD	Surgical (heart lung transplant)	Follow-up not mentioned
Sakamaki et. al.[5]	Primary PH	Surgical (reanastomosis)	Stable during 37 months of follow-up
Westaby et al.[7]	Secondary PH from VSD, Eisenmenger's syndrome	Surgical (vascular prosthesis)	Follow-up not discussed, stable on discharge on tenth day
Senbaklavaci et al.[12]	Primary PH	Surgical	Stable during 10 months of follow-up
Inayama et al.[2]	PH secondary to pulmonary thrombosis	Surgical	Follow-up not discussed, stable at discharge
Wunderbaldinger et al.[13]	Primary PH	Surgical	Follow-up not discussed
Lopez-Candales et al.[14]	Secondary PH from partially corrected pulmonary stenosis	Surgical	Follow-up not discussed, stable on discharge at 1 week
Khatchatourian and Vala[17]	Associated with aortic dissection	Surgical	Stable during 3 months of follow-up
Rosenson and Sutton[6]	Secondary PH from mitral stenosis	Management not discussed	Follow-up not discussed
Stern et al.[16]	Secondary PH from hypersensitivity pneumonitis	Management not discussed	Follow-up not discussed

Abbreviations: ACE, angiotensin-converting enzyme; CCB, calcium channel blockers; COPD, chronic obstructive pulmonary disease; PAD, pulmonary artery dissection; PH, pulmonary hypertension; VSD, ventricular septal defect.

should be used cautiously in patients taking sildenafil or similar agents for erectile dysfunction or pulmonary artery hypertension because of significant risk of cardiovascular collapse. The American Heart Association and American College of Cardiology both recommend that there should be a time gap of at least 24 hours between the last dose of sildenafil and the first dose of nitrates. Conservatively managed patients should be followed with interval CT scans[2,9,18] or echocardiography.[3,19] In addition, the underlying etiology should always be investigated to predict prognosis and recommend future management strategies.

The patient was offered surgical repair but she declined. She was managed conservatively with nitrates and beta-blockers and was pain free within 24 hours. Her BP was brought down to a systolic BP range of 130–140 mm Hg. A repeat CT scan of the chest at 1-month follow up was unchanged. The patient was doing well at 6-month follow-up.

Conclusions

PAD is an extremely rare cause of chest pain and a rare antemortem diagnosis. It is usually associated with underlying pulmonary hypertension. This case describes a patient with PAD in the absence of pulmonary hypertension. The patient was managed with conservative medical therapy and did well at 6-month follow-up. There are a total of 6 other case reports of patients with PAD managed conservatively, out of which 5 patients did well at follow-up and 1 patient died. More case reports and longer follow-up are needed to assess the effectiveness of conservative medical therapy in patients with PAD. To our knowledge, this is the first case report of idiopathic PAD diagnosed in a living patient and managed conservatively. This case also highlights better prognosis for patients with PAD without underlying pulmonary hypertension.

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