MAYO CLINIC

39-Year-Old Woman With Dyspnea and Chest Pain

Scott A. Helgeson, MD; Asha U. Nookala, MD; and Joseph L. Blackshear, MD

A 39-year-old woman was admitted to the hospital with a 7-month history of dyspnea that had worsened over the preceding 2 days, along with several months of pleuritic chest pain that radiated from the left side of the chest to the back. Her medical history was notable for atrial fibrillation (AF) treated with catheter ablation 10 months earlier and nonischemic congestive heart failure diagnosed at age 30 years at an outside institution. The patient did not have orthopnea, edema, cough, or weight gain. She had been treated previously with warfarin prophylaxis for AF, but this treatment was discontinued 2 months before presentation. Her family history was notable for pulmonary embolism in her father at age 40 years.

The patient had been hospitalized twice at other facilities in the preceding 2 months, the first hospitalization for hypoxemic respiratory failure. Work-up included chest radiography and computed tomography (CT), which revealed patchy areas of ground-glass opacities and bilateral consolidations. Echocardiography documented an ejection fraction of 63%, normal diastolic function, and an estimated mean pulmonary artery pressure of 24 mm Hg. A video-assisted thoracoscopic lung biopsy identified nonspecific interstitial pneumonitis. The patient received a 7-day course of meropenem and linezolid. During the second hospitalization, pulmonary infiltrates were treated with cefepime and levofloxacin.

On presentation to our facility, the patient's vital signs were normal, and physical examination findings were remarkable for an increased pulmonic second heart sound on cardiac auscultation and diffuse chest wall tenderness. No jugular venous distention was noted, nor were cardiac murmurs, right ventricular lift, peripheral edema, or stigmata of chronic liver disease or venous congestion.

Electrocardiography revealed sinus tachycardia (heart rate, 106 beats/min) with no other abnormalities. Chest radiography identified diffuse bilateral interstitial opacities with Kerley B lines suggestive of pulmonary edema. Echocardiography confirmed the findings of the outside study but revealed a prominent color jet emanating from the right superior pulmonary vein and accelerated diastolic flow velocity of greater than 1 m/s (reference range,[1] 0.47-0.54 ms) by Doppler. Laboratory testing revealed the following notable results (reference ranges provided in parentheses): hemoglobin, 11.2 g/dL (12.0-15.5 g/dL); white blood cell count, 20.4 × 10^9/L (3.5-10.5 × 10^9/L); segmented neutrophils, 92.5% (44.4%-70.9%); immature granulocytes, 1.1% (0.0%-3.0%); lymphocytes, 4.1% (17.8%-41.5%); quantitative D-dimer, 1.7 μg/mL (≤0.5 μg/mL); arterial pH, 7.43 (7.35-7.45); partial pressure of arterial carbon dioxide, 33.7 mm Hg (35-45 mm Hg); partial pressure of arterial oxygen, 63.3 mm Hg (80-100 mm Hg); glucose, 295 mg/dL (70-140 mg/dL); lactate, 2.2 mmol/L; B-type natriuretic peptide, 82 pg/mL (≤64 pg/mL); troponin T, <0.01 ng/mL (<0.01 ng/mL); and serum procalcitonin, <0.05 ng/mL (≤0.15 ng/mL).

1. Which *one* of the following is the *most likely* explanation for this patient's dyspnea?

a. Acute coronary syndrome
b. Constrictive pericarditis
c. Pulmonary vein stenosis (PVS)
d. Heart failure with preserved ejection fraction
e. Pulmonary artery hypertension (PAH)

Acute coronary syndromes typically occur with angina that is substernal, lasts a few minutes, occurs with exertion, and is relieved with rest or nitroglycerin. Acute coronary syndrome is diagnosed on the basis of clinical presentation, electrocardiographic findings, and troponin levels. Constrictive pericarditis is one cause of heart failure with a normal left ventricular ejection

See end of article for correct answers to questions.

Resident in Internal Medicine, Mayo School of Graduate Medical Education, Jacksonville, FL (S.A.H.); Resident in Internal Medicine, Mayo School of Graduate Medical Education, Rochester, MN (A.U.N.); Advisor to residents and Consultant in Cardiovascular Diseases, Mayo Clinic, Jacksonville, FL (J.L.B.).

fraction. Most cases are idiopathic, and AF is commonly associated. However, jugular venous distention, peripheral edema, and possibly ascites would be expected, as well as left atrial enlargement.

Pulmonary vein stenosis has been described as a complication arising after catheter ablation for AF,[2] with onset of clinical findings typically 3 to 6 months after ablation. Pulmonary vein stenosis may cause a pulmonary edema pattern on chest radiography, consolidation-like findings over the affected lung such as dullness to percussion, increased fremitus, whisper pectoriloquy, bronchial breath sounds, and crackles. Recurrent pneumonias are common with PVS, and hemoptysis may occur. Pulmonary artery hypertension may or may not develop secondary to the stenosis. There are no laboratory findings consistent with PVS.

The patient had normal echocardiographic findings including normal left ventricular diastolic function. Although she had a history of AF, which is often associated with heart failure with preserved ejection fraction, there was no evidence of underlying diastolic dysfunction on echocardiography. Pulmonary artery hypertension presents nonspecifically with exertional dyspnea, fatigue, and occasionally syncope that develop over the course of years. Once PAH becomes severe, right ventricular failure develops and exertional chest pain, syncope, edema, ascites, and pleural effusion may start to appear. Physical examination usually elicits an increased intensity of the pulmonic second heart sound. Echocardiography will identify a mean pulmonary artery pressure of more than 25 mm Hg.

Echocardiography confirmed the findings of the outside study but also revealed a prominent color jet emanating from the right superior pulmonary vein and accelerated diastolic flow velocity of more than 1 ms by Doppler, which is seen in PVS. This finding, in conjunction with knowledge of the potential complications from the prior ablation procedure, suggested a diagnosis of PVS.

2. At this time, which *one* of the following is the *most appropriate* test to establish a diagnosis of PVS?

a. Transesophageal echocardiography
b. Bronchoscopy
c. Ventilation-perfusion scan and venous phase–gated CT angiography
d. Oxygen consumption exercise study
e. Right-sided heart catheterization with nitric oxide challenge

Transesophageal echocardiography is able to visualize all the pulmonary veins in 96% of patients and may reveal evidence supporting a diagnosis of PVS, but image interpretation is highly dependent on the experience of the physician, and precise anatomic detail such as lumen diameter and length of stenosis are not definable by transesophageal echocardiography.[3] Bronchoscopy is unlikely to establish a tissue diagnosis in a patient with pulmonary infiltrates who already had an open lung biopsy.

Venous phase–gated CT angiography is the test most likely to define the precise anatomy of the junction of the 4 pulmonary veins to the left atrium. The venous phase detail is important because standard pulmonary embolism protocol–gated CT angiography or nongated CT scanning would lack sufficient spatial resolution in the pulmonary veins for this purpose. The physiologic consequences of PVS would best be appreciated with radionuclide ventilation-perfusion lung scanning for evidence of perfusion defects that match the PVS.

An exercise test with measured oxygen consumption is an excellent means of assessing maximum cardiac output in patients with known cardiac disorders. Although this test has value in distinguishing cardiogenic dyspnea from dyspnea due to lung disease, it would not aid in making a precise diagnosis in this instance. Right-sided heart catheterization accurately measures pulmonary artery pressures, pulmonary vascular resistance, and pulmonary capillary wedge pressure, and nitric oxide inhalation assesses pulmonary vascular responsiveness, which aids in choosing treatment in patients with PAH. This test allows detailed visualization of the pulmonary veins but is an invasive test. This test should be performed but is not the best test at this time.

Our patient underwent venous phase CT angiography, which revealed occlusion of the left superior pulmonary vein and high-grade stenosis of the left inferior pulmonary vein and both right pulmonary veins. Ventilation-perfusion scanning documented absent perfusion to the left lung. On the basis of the clinical presentation and the imaging results, symptomatic PVS

Mayo Clin Proc. ■ XXX 2016;■■(■):1-5 ■ http://dx.doi.org/10.1016/j.mayocp.2015.12.020
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