# A 46-Year-Old Man With Dyspnea, Dysphagia, and Pulmonary Hypertension*

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***(CHEST 2008; 133:812–815)***

A 46-year-old man presented to the emergency department with a 2-day history of dyspnea on exertion, orthopnea, productive cough, and pleuritic chest pain. He had no fever or chills. His medical history was significant for type II diabetes mellitus and acute myocardial infarction. Current medications included aspirin, enalapril, atorvastatin, metoprolol XL, and metformin. He had a 40-pack-year history of cigarette smoking. He had no history of alcohol or IV drug use. He was married and had no risk factors for HIV infection. A review of systems revealed decreased appetite, a 25-lb weight loss over 2 weeks, and intermittent dysphagia for 2 weeks. He had no skin lesions, skin tightening, heart burn, arthralgias, hematuria, or history of cytopenias.

*Physical Examination*

Vital signs included a pulse rate of 105 beats/min, a BP of 101/77 mm Hg, a respiratory rate of 14 breaths/min, a temperature of 97.3°F, and an $O_2$ saturation of 95% while breathing oxygen at 3 L/min. He had diminished breath sounds at the lung bases and a few intermittent bilateral wheezes.

*Laboratory and Radiologic Studies*

Routine laboratory findings included the following: hemoglobin concentration, 17.1 g/dL; hematocrit, 50.7%; WBC count, 12,500 cells/μL; platelet count, 395,000 cells/μL; normal WBC differential count; sodium concentration, 133 mmol/L; potassium concentration, 4.8 mmol/L; chloride concentration, 97 mmol/L; $CO_2$ level, 19 mmol/L; BUN concentration, 14 mg/dL; and creatinine concentration, 1.0 mg/dL. Arterial blood gas measurements revealed a pH of 7.46, a $P_{CO_2}$ of 20 mm Hg, and a $P_{O_2}$ of 51 mm Hg on room air. Troponin T levels were $< 0.01$ ng/mL on three sequential measurements. The pro-brain natriuretic peptide concentration was 741 pg/mL (normal concentra-

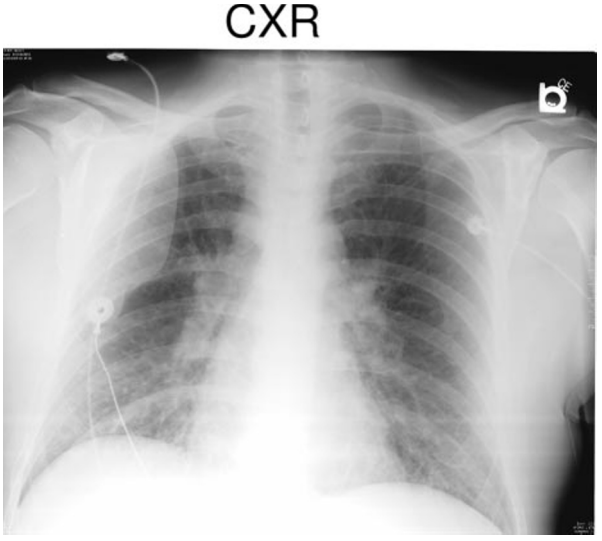


FIGURE 1. Hospital admission chest radiograph. The density in the right upper thorax is a cardiac monitor pad.

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The authors have reported to the ACCP that no significant conflicts of interest exist with any companies/organizations whose products or services may be discussed in this article.

Manuscript received July 29, 2007; revision accepted August 27, 2007.



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**DOI: 10.1378/chest.07-1871**

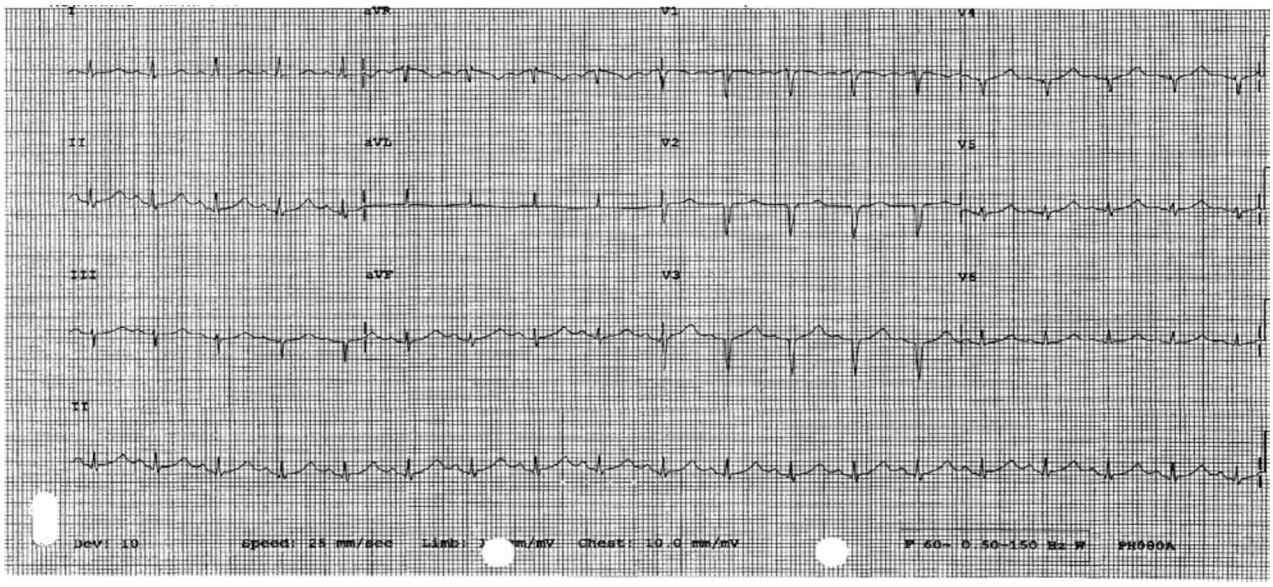


FIGURE 2. Hospital admission ECG.

tion, 5 to 400 pg/mL). The d-dimer concentration was > 1,000 ng/L (normal concentration, 68 to 494 ng/L). Hospital admission chest radiograph and ECG are shown in Figures 1 and 2, respectively. The findings of a CT scan of the chest were negative for pulmonary emboli but did reveal a large pulmonary artery outflow tract. Multiple hilar and mediastinal lymph nodes with a mass in the right retrocarinal space (3.3 × 2.7 cm) were present. An esophagram revealed slight narrowing of the distal thoracic esophagus and minimal gastroesophageal reflux. Systolic pulmonary artery pressures were estimated to be 93 to 98 mm Hg by transthoracic echocardiogram. Pulmonary artery pressures were normal in a study performed 4 months earlier. Cardiac catheterization revealed a pulmonary artery pressure of 64/29 mm Hg, a pulmonary capillary wedge pressure of 24 mm Hg, a right atrium pressure of 4 mm Hg, a right ventricle pressure of 61/3 mm Hg, and a left ventricle pressure of 92/4 mm Hg. No pulmonary emboli were found.

*What is the cause of this patient's pulmonary hypertension?*