

Abstract:

This is the case of a 13-year-old adolescent girl who presented to the emergency department and was diagnosed with severe pulmonary hypertension. In the intensive care unit, she underwent a comprehensive evaluation for the underlying etiology of her pulmonary hypertension and was determined to have pulmonary venoocclusive disease, a rare disorder of pulmonary venous fibrosis and thrombosis.

Keywords:

pulmonary hypertension; pulmonary venoocclusive disease; chest pain; dyspnea; right heart failure

A 13-Year-Old With Fatigue, Chest Pain, and Dyspnea

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A 13-year-old African American adolescent girl presented to the emergency department (ED) with a 2-month history of fatigue, shortness of breath, and chest pain. She had initially noted these symptoms while climbing the staircase at her school. In the ED, she described her chest pain as sharp, midsternal, and nonradiating and stated that it occurred with activity and at rest. The duration of these episodes of chest pain ranged from 5 minutes to an hour, with multiple episodes per day that were increasing in frequency and severity at the time of presentation. For the past 2 months, she had also experienced constant, dull epigastric pain, right flank pain, and back pain. In addition, she reported constipation.

Approximately 1 1/2 months before presentation, she had been evaluated in the ED of another hospital and had been diagnosed with a viral illness and gastritis. A chest x-ray and electrocardiogram (EKG) had been performed, and her mother understood the results of these to be normal. She had been prescribed ranitidine and a laxative. Shortly after this first ED visit, she had a syncopal episode while straining on the toilet. She was then admitted to the same hospital where she had first sought care. An etiology of her symptoms was not identified during that admission. Her mother reported that a cardiac murmur had been noted and that she had been discharged with 2-month follow-up visit with a cardiologist.

In the 6-week interval since that first ED evaluation, her fatigue and shortness of breath had progressed; she was unable to walk across a room without feeling tired. As a result of her fatigue, she had been unable to attend school for the past week. On the day of

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presentation, the patient had a 5-minute episode of pallor and perioral cyanosis that had been observed by her mother. On that day, her mother had also noted that the patient appeared to be swollen in her face, hands, and legs.

The patient's past medical history was only significant for an episode of "wheezing" at the age of 2 years. Her review of systems was negative for fever and positive for an occasional nonproductive cough. Her initial vital signs included a temperature of 36.8°C, a heart rate of 120 beats per minute, a respiratory rate of 24 breaths per minute, and a blood pressure of 92/68 mm Hg. Her oxygen saturation was 96% on room air. She had slight periorbital fullness without discoloration. Extraocular movements were normal. Her heart sounds were notable for a loud S1, S2, and an S3 gallop. No murmurs were appreciated. Her lungs were clear, without crackles or wheezes. Her abdomen was tender to palpation in the right upper quadrant and epigastric region. The liver was palpated 3 cm beneath the right costalmargin. Her right flank was tender. Her extremities were warm and pink, with nonpitting edema of her hands and feet. There was 1+ pitting edema of her pretibial skin.

Laboratory workup in the ED included a complete blood count, complete metabolic panel, and a urinalysis. An EKG and chest x-ray were also performed. The white blood cell count was 7610/ μ L (4500-13500/ μ L) with a normal differential. Her hemoglobin, hematocrit, and platelets were within reference range. A basic metabolic panel was notable for slightly low serum bicarbonate of 18 mEq/L and calcium of 8.2 mg/dL (8.8-10.8 mg/dL). Liver function tests were slightly elevated, with an aspartate aminotransferase of 55 IU/L and alanine aminotransferase of 56 IU/L. A urinalysis revealed trace protein. A chest x-ray showed poorly vascularized peripheral lung fields, an increased cardiothoracic ratio, a prominent right heart border, and a prominent main pulmonary artery segment (Figure 1). An EKG demonstrated right axis deviation, right ventricular hypertrophy with a strain pattern, and a QTc of 475 milliseconds (Figure 2).

DIFFERENTIAL DIAGNOSIS

In general, the differential diagnosis for all causes of chest pain is extensive and includes the following categories: musculoskeletal/neural, tracheobronchial, pleural, esophageal, cardiac, and psychogenic (anxiety, hyperventilation).^{1,2}

- Musculoskeletal causes include strain of chest wall musculature or inflammation of costo-

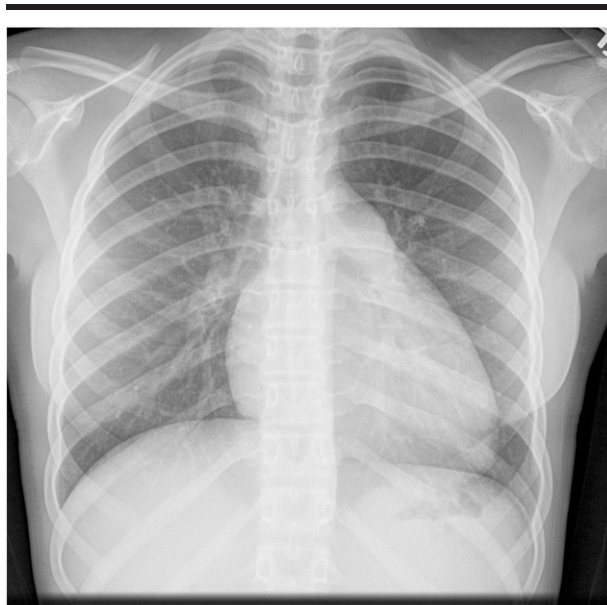


Figure 1. Chest x-ray with borderline cardiomegaly, poorly vascularized lungs, a prominent pulmonary artery, and a pronounced right heart border.

chondral junctions (costochondritis). Also incorporated in this category are contusions, lacerations, or infections of muscle, breast tissue, or bone. Rib fractures are another relatively common etiology.

- Neurally mediated chest pain includes neuritis or radiculitis due to toxins, trauma, or infection.
- Tracheobronchial and pulmonary chest pain incorporate pain due to foreign bodies, tracheitis, bronchitis, pneumonia, or asthma.
- Pleural-related chest pain may arise from trauma, pleural inflammation (viral, mycobacterial, or autoimmune), pneumonia, pleural effusion, pulmonary embolus, vasoocclusive crisis, cholecystitis, pneumothorax, or pneumomediastinum. Mediastinal masses are included in this subset.
- Gastrointestinal causes of chest pain include esophageal foreign body, caustic ingestion, gastritis, gastroesophageal reflux, achalasia, esophagitis, and referred pain from cholecystitis or pancreatitis.
- Cardiac sources include coronary vasospasm, anomalous left coronary artery, coronary aneurysm (Kawasaki disease), obstructive cardiac disease, pericardial effusions, pericarditis, arrhythmia, myocarditis, and aortic aneurysm.
- Precordial catch syndrome is a common but underrecognized cause. Chest pain is characteristically sharp, is well localized, and lasts

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