Abnormal pulmonary density discovered on routine chest radiograph

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A routine chest radiograph of a 69-year-old woman revealed an abnormal density in the left lung. There was no history of a previous illness. Physical examination showed a single tel-

angiectasia on the tongue. Radiographic and imaging studies are shown below (Figures 1–4).

For diagnosis and discussion, see the following page.

Figure 1. Posteroanterior radiograph of the chest demonstrates an abnormal density (arrows) in the inferior portion of the left lung.

Figure 2. Computed tomography (CT) of the chest (lung window) shows a lobulated mass (arrows) that corresponds to the abnormal pulmonary density.

Figure 3. Enhanced CT image of the liver demonstrates an irregular vascular liver mass (arrowheads) and innumerable small vascular liver masses (arrows).

Figure 4. Enhanced CT image of the liver shows enlargement of the hepatic artery (arrows). Innumerable small, irregular vascular liver lesions are again seen (arrowheads).

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DIAGNOSIS: Hereditary hemorrhagic telangiectasia (HHT).

DISCUSSION

HHT, or Osler-Weber-Rendu disease, is an autosomal-dominant disorder characterized by multiple mucocutaneous and visceral vascular abnormalities consisting of thin-walled, dilated vascular channels with arteriovenous malformations (AVM). Any organ can be involved. The lesions affect the mucocutaneous tissue 78% of the time; the gastrointestinal tract, 44%; the liver, 30%; the brain, 28%; the lungs, 15% to 20%; and the spine, 8% (1). Aneurysms involving any size vessel may also occur (2). In the current case, the patient presented with a pulmonary AVM. Thirty percent to 88% of pulmonary AVMs are associated with HHT, whereas 15% to 20% of patients with HHT have pulmonary AVMs. Other causes of pulmonary AVMs include trauma, cirrhosis (hepatogenic pulmonary angiodysplasia), and infection (3).

The prevalence of the disorder is reportedly between 1 in 50,000 to 1 in 100,000 (4). Patients with the disorder will manifest symptoms by the third or fourth decade (3). The defect reportedly involves the endoglin gene. Endoglin is a glycoprotein found on the endothelial cells of arterioles, venules, and capillaries (5).

Patients with HHT manifest a multitude of symptoms depending on the size and location of their lesions. The disorder can be disfiguring because of the numerous mucocutaneous telangiectasias involving the face, lips, tongue, ears, nasal mucosa, and hands. Mucosal lesions can cause episodes of epistaxis in up to 78% of patients (3). Epistaxis combined with chronic gastrointestinal bleeding can be severe enough to cause significant iron deficiency anemia (6). Dyspnea on exertion is a common manifestation of the disorder. Dyspneic symptoms are explained by large and/or multiple pulmonary AVMs causing right-to-left shunting and subsequent hypoxemia. Other factors involved include chronic anemia and high-output cardiac failure. Additional cardiopulmonary symptoms include hemoptysis secondary to rupture of an AVM into a bronchus and hemothorax secondary to rupture of a subpleural AVM (3).

Hepatic involvement is common but usually causes cardiovascular symptoms when symptomatic. Hepatic AVMs cause cardiovascular symptoms when the intrahepatic shunting is >20% of total cardiac output, leading to heart failure and pulmonary hypertension (6). Primary liver manifestations are less common, but arterioportal shunts can lead to fibrosis and cirrhosis (1).

The most serious sequelae of HHT involve the central nervous system. Nearly half the cases with neurologic complications occur secondary to pulmonary AVMs. Cerebrovascular accidents, transient ischemic attacks, and brain abscesses are potential complications of paradoxical emboli. Venous infarcts can occur in patients with polycythemia secondary to chronic hypoxemia.

AVMs and intracranial aneurysms may also lead to intracranial hemorrhage (2).

AVMs can be imaged using many modalities. Radiography is mostly useful in the evaluation of pulmonary AVMs. An extensive differential diagnosis of a solitary pulmonary nodule includes AVMs. The correct diagnosis is suggested when there are multiple lobulated pulmonary lesions and when a feeding artery and a draining vein are visible. These malformations occur commonly in the lower lobes and may change in size with Valsalva's maneuver or patient positioning.

Doppler sonography allows analysis of the flow patterns of hepatic vascular malformations. Findings include dilatation and increased tortuosity of the hepatic artery (1). Arterioportal and arteriovenous shunting alter the waveforms of the portal and hepatic veins. Portal shunting causes pulsatility and/or reversal of the normally smooth hepatopedal flow (4). The normal hepatic venous waveform usually varies with the cardiac cycle and the phase of respiration. Arteriovenous shunting creates a more pulsatile Doppler waveform due to the lack of an intervening capillary bed.

CT and magnetic resonance imaging (MRI) are useful for identifying multiple lesions and further defining the anatomy. Dynamic contrast-enhanced CT and magnetic resonance angiography (MRA) are able to demonstrate the feeding artery and the draining vein when the malformation is located in the lungs, liver, or brain. CT and MRI are also able to show the complications of multiple AVMs including cirrhosis, intracranial hemorrhage, and brain abscesses. MRI and MRA have the added benefits of multiplanar capability, the lack of iodinated contrast, and the lack of ionizing radiation.

Angiography is reserved for preoperative evaluation and for treatment. The treatment of choice for patients with multiple AVMs is transcatheter embolization with coils or detachable occlusion balloons. In addition to embolization, liver transplantation is used to treat patients with severe cardiac and hepatic manifestations when medical therapy is ineffective (6).

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