A 29-year-old white man presented with a 2-day history of progressive dyspnea and chest tightness. He was from Minneapolis, MN (altitude, 0 m), and had arrived in Winter Park, CO (altitude, 2,750 m), 1 day before his symptoms began. One week prior to travel, he had symptoms of an upper respiratory tract infection including fatigue, diaphoresis, fever, and productive cough. He was an otherwise healthy non-smoker who reported having childhood asthma but had experienced no symptoms during adulthood.

A physical examination revealed an uncomfortable man breathing 32 times per minute, who was in moderate respiratory distress. The patient’s pulse oximetry while breathing room air was 62%, which improved to 95% with the addition of supplemental oxygen via a non-rebreather mask. A chest examination demonstrated symmetrical expansion, bibasilar rales, and no wheezes. The results of a cardiac examination were normal, and the P₂ sound was not

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Figure 1. Posteroanterior chest radiograph on presentation.

Figure 2. Lateral chest radiograph on presentation.
accentuated. Pertinent laboratory studies indicated the following: normal leukocyte count; hematocrit, 52%; and lactate dehydrogenase level, 587 U/dL. The chest radiograph demonstrated a left perihilar infiltrate and herniation of the left lung through the anterior mediastinum (Fig 1, 2). A CT scan of the chest was performed (Fig 3, 4).

What is the diagnosis?
Diagnosis: Unilateral pulmonary artery agenesis with high-altitude pulmonary edema in the contralateral lung

Discussion

First described in 1868, unilateral pulmonary artery agenesis (UPAA) remains a rare developmental anomaly. It is also referred to as proximal interruption of the pulmonary artery, because in some cases surgical dissection reveals an incompletely developed pulmonary artery at the level of the hilum. Agenesis of the right pulmonary artery is due to a lack of development of the right sixth aortic arch. Intrapulmonary vessels and the distal portion of the right pulmonary trunk often develop normally and receive their blood supply from bronchial vessels. This results in a small hypovascular right lung. The absence of the right pulmonary artery in a patient occurs with greater frequency than that of the left pulmonary artery and may be associated with the presence of patent ductus arteriosus. The absence of the left pulmonary artery often is associated with other cardiovascular abnormalities such as tetralogy of Fallot, right aortic arch, patent ductus arteriosus, and septal defects.

The clinical presentation of right UPAA is variable and often is detected by plain chest radiographs that have been taken for other reasons. Right UPAA often occurs as an isolated anomaly, with approximately 30% of patients remaining asymptomatic throughout their lives. Hemothysis occurs in up to 10% of patients secondary to either hypertrophied bronchial collateral vessels or peripheral arteriovenous fistulas. Other patients may present with recurrent infections, dyspnea on exertion, and congestive heart failure. Left UPAA is usually recognized in childhood due to the high frequency of associated cardiovascular abnormalities. UPAA also is associated with inordinate shortness of breath and exercise limitation when compared to the condition of adolescent peers.

The diagnosis of UPAA often is based on medical history, physical examination findings, and the results of chest radiographs. As demonstrated here, the chest radiograph frequently reveals hypovascularity and a reduced size of the right lung with herniation of the left lung through the anterior mediastinum. Other findings include a small hilum, hyperlucency, and a mediastinal shift toward the affected lung. Additional confirmatory studies include angiography, ventilation/perfusion scanning, CT scan, and MRI.

High-altitude pulmonary edema has been reported in patients with an absence of the right pulmonary artery. In 1980, Hackett et al reported four cases of patients with right-sided UPAA, all of whom developed high-altitude pulmonary edema at moderate altitudes in Colorado. The absence of the right pulmonary artery was established at autopsy in one case, by angiography in two cases, and by perfusion lung scanning in the fourth case. Theoretically, the reduced overall size of the pulmonary vascular bed leads to a higher pulmonary artery pressure in response to hypoxic vasoconstriction at any given altitude. This results in overperfusion of the affected lung and the development of pulmonary edema.

Several other entities should be considered in the differential diagnosis of UPAA. Swyer-James (also known as Swyer-James-McLeod) syndrome is the result of childhood bronchiolitis that leads to lobar or segmental emphysema. Significant air trapping is seen in the affected lung, whereas in patients with UPAA, the affected lung demonstrates relative underventilation when compared to the hypertrophied contralateral lung. Chronic venous thromboembolic disease also can mimic UPAA. Moser et al described three subjects with a presumed diagnosis of UPAA who were ultimately found to have chronic thromboembolic occlusion. The clinical presentation may be similar, with progressive dyspnea and hemoptysis, and the chest radiograph findings may be similar, showing a relative absence of the pulmonary artery shadow, decreased lung volume, and hypovascularity on the affected side. However, physical signs of pulmonary hypertension and lower extremity venous obstruction are more likely to be seen in patients with chronic thromboembolic occlusion. Ventilation-perfusion scanning, CT scanning, and pulmonary arteriography also may help to differentiate the two entities.

While most patients with left UPAA undergo corrective surgery for associated cardiovascular abnormalities, surgery is generally not indicated for patients with right UPAA. However, selected cases with recurrent hemoptysis or infections may benefit from surgical revascularization or pneumonectomy. Although no formal recommendations exist, the avoidance of high altitude should be strongly considered in patients with UPAA. Symptoms may occur at elevations of 6,000 to 7,000 feet, and deaths have been reported at these modest altitudes. If the avoidance of travel to high altitudes is not possible, the use of supplemental oxygen or prophylaxis with nifedipine, 30 mg twice daily, should be considered. Given the potentially life-threatening complications that can result from travel to high altitudes, a heightened clinical awareness and the prompt diagnosis of UPAA is important.
REFERENCES


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