High-Altitude Pulmonary Edema in Partial Anomalous Pulmonary Venous Connection of Drainage with Intact Atrial Septum*

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Partial anomalous pulmonary venous drainage may be the cause of hypoxemia and overperfusion of the lungs and therefore an important factor in the pathogenesis of high-altitude pulmonary edema (HAPE).

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ASD = atrial septal defect; HAPE = high-altitude pulmonary edema; PAPVD = partial anomalous pulmonary venous drainage

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High altitude pulmonary edema (HAPE) has been a recognized entity since 1960. Its underlying pathophysiology remains unclear. The occurrence of partial anomalous pulmonary venous return (PAPVD) without an associated atrial septal defect (ASD) is uncommon. We describe one patient with PAPVD who experienced two episodes of HAPE at moderate altitude. The question is if the hemodynamic alterations in this cardiac abnormality play a role in the pathogenesis of HAPE.

**Case Report**

A 29-year-old mountaineer living in Holland has been climbing regularly in mountains of 3,000 to 7,000 m for 12 years. In December 1989, he made a trip to the Swiss mountains. In three days, he ascended to 3,800 m. At night, he developed shortness of breath and a nonproductive cough. He descended to 2,400 m and was taken to a hospital. On admission, he was cyanotic. The blood pressure was 150/100 mm Hg, and the pulse was 90 beats per minute. Rales were present over both lung bases. A grade 3/6 mid-late systolic crescendo-decrescendo murmur and an accentuated pulmonary second sound were heard along the left sternal border. Blood gas analysis showed respiratory alkalosis. An x-ray film of the chest revealed pulmonary edema, especially in the left lower lobe, and prominent dilated main pulmonary arteries (Fig 1).

The ECG showed sinus rhythm with signs of right atrial enlargement. The patient was treated with nifedipine (20 mg). Because an ASD or an anomalous pulmonary venous return was considered, further examination was performed. On the bicycle ergometer a significant decrease in partial oxygen pressure was measured: 93 mm Hg to 77 mm Hg. Cardiac catheterization showed an intact atrial septum but anomalous drainage of the upper and middle lobe vein of the right lung into the superior vena cava at the junction of the innominate vein and the superior vena cava (Fig 2).

**Figure 1.** Chest x-ray film showing HAPE.

**Figure 2.** Cardiac angiogram showing anomalous pulmonary venous drainage of right upper and middle lobe vein into right atrium (RA) and of right lower lobe vein into left atrium (LA). PVD, Pulmonary vein dexter.
with the right atrium (Fig 2). The pulmonary-to-systemic blood flow ratio was 2.6:1. The systolic pulmonary artery pressure and the pulmonary vascular resistance were normal. On his next expedition the patient experienced a second episode of HAPe at 6,400 m, while receiving acetazolamide. In December 1990, reconstruction of the pulmonary anomaly was performed. The angiographic findings were confirmed.

DISCUSSION

The pathogenesis of HAPe is still incompletely understood. Hypoxemia appears to be of paramount importance. At high altitude the partial pressure of oxygen in the inspiratory air decreases, leading to pulmonary vasoconstriction and pulmonary hypertension. Exercise leads to a further decrease in arterial saturation, due to the shortened transit time of blood in the alveolar capillaries. The capillary wedge pressure is normal, in contrast to the elevated pulmonary artery pressure. The arteriolar vasoconstriction is probably not uniform throughout the lungs.

Subjects with an increased vasoconstrictor response on breathing a hypoxic gas mixture appear to be at increased risk to develop HAPe, and show an uneven lung perfusion on radionuclide scan. In subjects with congenital absence of the right pulmonary artery, HAPe was seen only on the left side, illustrating the supposition that overperfusion of the lung is important in the pathogenesis of HAPe. The edema fluid has a high protein content, suggesting that it is a form of permeability edema. Alpert et al found in 14 patients with right PAPVD without ASD a higher blood flow through anomalously draining lobes of the lung than through normally draining lobes. The pulmonary vascular resistance was approximately the same for normal and anomalous lung segments. Pulmonary artery pressure was identical for both lungs.

In our patient a pulmonary-to-systemic blood flow ratio of 2.6:1 was found, leading to an increased blood flow through the lungs during exercise. He suffered two episodes of HAPe, one at a relatively low altitude, which suggests that PAPVD was a significant factor in the pathogenesis of HAPe. There are two possible explanations for his increased susceptibility for HAPe. First, arterial undersaturation was probably more pronounced at altitude compared to healthy individuals; exercise on the ergometer at sea level showed a remarkable decrease in saturation. Secondly, pulmonary overperfusion occurred because of PAPVD. This case history underlines the importance of hypoxemia and overperfusion of the lungs in the pathogenesis of HAPe.

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Massive Hemothorax Associated with Intrathoracic Extramedullary Hematopoiesis Involving the Pleura*

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Intrathoracic extramedullary hematopoiesis rarely involves the pleura and is usually asymptomatic. We report a 73-year-old woman with myelofibrosis who had pleural involvement with extramedullary hematopoietic tissue that produced a massive hemothorax. Before the diagnosis of extramedullary hematopoietic tissue was established, sclerosis with tetracycline was attempted, which accelerated pleural bleeding and required surgical evacuation. The bleeding was ultimately controlled by low-dose radiation therapy.

Extramedullary hematopoiesis, the formation of apparently normal blood cells outside the confines of the bone marrow, usually occurs as a compensatory response either to chronic hemolytic conditions or bone marrow replacement because of tumor or fibrotic tissue. The heterotopic marrow is usually microscopic and most commonly involves the spleen, liver, and lymph nodes, but can also be seen in adipose tissue and in tissue of the adrenal gland, kidney, thymus, peripheral nerve, breast, cartilage, broad ligament, pleura, retroperitoneum, epididymis, and epididymis. At times, the heterotopic site can be large and can present as a tumor mass or masses.

Intrathoracic extramedullary hematopoietic tissue most commonly occurs in the posteroinferior mediastinum, but has been reported in the anterior mediastinum and pleura. Pleural involvement is usually microscopic and asymptomatic and is noted only at autopsy. We present the case of a patient with intrathoracic extramedullary hematopoiesis involving the pleura that presented as a bloody pleural effusion of unclear cause and became a hemothorax after sclerotherapy with tetracycline.

CASE REPORT

A 73-year-old woman with a history of myelofibrosis diagnosed 7 years earlier presented with dyspnea and a right-sided pleural effusion. Sanguineous fluid (1,000 ml) was removed, after which her symptoms improved. The pleural fluid was an exudate and had an

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