Serpiginous Lung Densities*

Marshall E. Bein, M.D.; **Michael J. Kelley, M.D.; and William E. Hellenbrand, M.D.†

A 46-year-old Puerto Rican woman was admitted for evaluation of an abnormality noted on routine chest roentgenogram. She gave a history of recent dyspnea on exertion, but had had no cardiac or pulmonary disease. There was a 40 to 60 pack-year history of cigarette smoking. Except for bibasilar rales, physical examination was normal. All routine laboratory data, including electrocardiogram, gave normal findings. Arterial blood gas studies on room air were pH 7.40, Po2 72 mm Hg, and Pco2 35 mm Hg. Pulmonary function tests were consistent with moderate obstructive pulmonary disease.

Chest roentgenograms in the PA and lateral projections are shown in Figures 1 and 2.

*From the Departments of Diagnostic Radiology and Pediatrics, Yale University School of Medicine and Yale-New Haven Hospital, New Haven.
**Winchester Fellow in Diagnostic Radiology of the Chest.
†U.S. Public Health Service Fellow, supported by Training Grant NIH (No. 5-T01-AD-00177).
Reprint requests: Dr. Kelley, Department of Diagnostic Radiology, Yale University School of Medicine, New Haven 06510

Figure 1

Figure 2
Diagnosis: Pulmonary varices

Figures 1 and 2 demonstrate multiple serpiginous tubular densities in the right middle and lower lung zones, contiguous with the posterior heart border. Chest tomography confirmed the vascular nature of the densities and demonstrated a small right descending pulmonary artery. Ventilation-perfusion scan was consistent with moderate air trapping at the lung bases. Cardiac catheterization yielded normal oxygen saturation, right heart pressures, and right pulmonary artery wedge pressure. Hydrogen electrode study in the right atrium and main pulmonary artery revealed no evidence of left-to-right shunt.

The pulmonary arteriogram (Fig 3 and 4) demonstrates diminution in size and stasis of flow in the lower lobe pulmonary arteries which, with the normal wedge pressure, is consistent with obstructive pulmonary disease. The levophase (Fig 5) shows: (1) opacification of dilated, tortuous veins in the right middle and lower lung zones, (2) normal appearance time of the two right and left pulmonary veins before their entry into the left atrium, and (3) no opacification of right heart structures. The angiographic findings are diagnostic of pulmonary varices.

The plain film differential diagnosis of serpiginous tubular densities in the lungs includes pulmonary arteriovenous malformation and partial anomalous pulmonary venous return, both of which may be differentiated by cardiac catheterization and angiocardiography. Pulmonary arteriovenous fistula is excluded in the present case by the absence of enlarged pulmonary arteries and early draining veins. Anomalous venous return is ruled out by normal oxygen saturation and normal hydrogen electrode study.

A pulmonary varix is a localized enlargement of a portion of a pulmonary vein that enters the left atrium normally. Embryologically, it probably occurs when the lung changes from splanchnic to pulmonary venous drainage. Possible theories concerning etiology include localized weakness of the venous wall and incorporation of the primitive splanchnic vein into the pulmonary venous system.

Although of uncertain etiology, pulmonary varices may not be related to other cardiopulmonary diseases, and their existence in patients with cardiac disease may be purely coincidental. A review of the literature reveals that heart disease, acquired or congenital, is the most common abnormality associated with pulmonary varices. Of 14 patients with associated heart disease, five had mitral insufficiency and five had mitral stenosis. Pulmonary venous hypertension in the patients with mitral disease may be responsible for enlargement of the pulmonary varices. Incidental associations of pulmonary varices with pulmonary tuberculosis, systemic hypertension, left upper lobe emphysema, intracranial aneurysm, and hyperparathyroidism have been reported.

As an isolated entity, pulmonary varices are usually unassociated with clinical signs and symptoms. Rare complications include rupture and systemic embolization. Treatment is unnecessary in asymptomatic patients.

Varices are most commonly located in the right lower and middle lobes. In the absence of coincidental left atrial hypertension, they remain unchanged in size over many years of observation. Definitive diagnosis requires pulmonary arteriography. The findings consist of a normal pulmonary arterial phase with opacification of the dilated tortuous veins in the levophase at the same time as the normal veins. Drainage is into the left atrium with delayed emptying of the varix.

REFERENCES