A 51-year-old woman was admitted to our hospital because of shortness of breath. Her mean pulmonary artery pressure was 52 mm Hg, and idiopathic pulmonary artery hypertension was diagnosed. She received beraprost sodium, imidapril, warfarin, and home-based oxygen therapy. At 67 years of age, dyspnea on effort worsened to New York Heart Association functional class III. The plasma level of brain natriuretic peptide was 57 pg/mL. Transthoracic echocardiography showed an enlarged right ventricle, severe tricuspid regurgitation, and a large pericardial effusion. At 68 years of age, she was urgently admitted to our hospital because of severe dyspnea at rest. A chest x-ray showed bilateral pulmonary artery enlargement (Figure 1).Computed tomography of the chest revealed that both main bronchi were compressed by the pulmonary artery aneurysms, with no mural thrombus in the pulmonary arteries (Figure 2). The diameters of the right and left main pulmonary arteries were 69.8 and 55.5 mm, respectively. Bronchoscopy during mechanical ventilation confirmed nearly complete extrinsic compression of the left and right main stem bronchi without positive airway pressure (Figure 3). On hospital day 3, continuous administration of epoprostenol was begun. She received continuous positive airway pressure by mechanical ventilation. Because the pulmonary hypertension had been worsening, treatment with imatinib (50 mg/d) was started on hospital day 69. However, the patient died of recurrent obstructive pneumonia 12 months after admission, and autopsy was conducted.

Histopathologically, pulmonary arteries/arterioles revealed dilatation, and alveolar walls were thickened with increased capillaries, deposition of hemosiderin, and loose fibrosis (Figure 4a and 4b). However, severe concentric medial hypertrophy, intimal fibrous thickening, and plexiform lesion, which are characteristic findings of idiopathic pulmonary artery hypertension, were not found in the pulmonary arteries/arterioles (Figure 4a and 4b). Normal pulmonary veins in the thickened interlobular septa mostly disappeared, and capillaries increased instead (Figure 4c and 4d).

The pulmonary veins might have been destroyed during the prolonged disease course. Enlarged bronchial veins (Figure 5) probably served as collateral vessels of the occluded pulmonary veins. On reviewing the computed tomography scans obtained at the patient’s initial admission, we found ground-glass opacities and smooth thickened intralobular septa, which are characteristic features of pulmonary veno-occlusive disease (PVOD; Figure 6).

Discussion

PVOD is a rare cause of pulmonary hypertension. The origin of PVOD is unknown. It might represent an idiopathic process or a common pathological response to various injurious stimuli.1 The symptoms of PVOD resemble those of pulmonary hypertension resulting from other causes and include progressive dyspnea on exertion, nonproductive cough, and edema of the lower extremities. However, medical treatment of primary pulmonary hypertension with prostacyclin can be fatal in patients with PVOD, and an early diagnosis is essential before therapy. PVOD is difficult to diagnose on routine
examinations (eg, blood tests, chest radiography, ECG, echocardiography, Swan-Ganz catheterization). High-resolution computed tomography is essential for the diagnosis of PVOD because a lung biopsy can worsen respiratory status.\(^2\) The presence of ground-glass opacities (particularly with a centrilobular distribution), septal lines, and adenopathy are indicative of PVOD.\(^3\) In patients with PVOD, the interlobular septal veins are occluded; the capillary network is secondarily dilated, forming multiple vascular loops; the lymphatic channels are enlarged; and the interlobular septum is edematous.\(^2\) The only curative therapy is lung transplantation.

In our patient, PVOD was definitively diagnosed at autopsy, although characteristic computed tomography findings had already developed (Figure 6). She survived for >15 years after initial admission, despite the poor prognosis of PVOD. To the best of our knowledge, this patient is the longest PVOD survivor.

Prolonged survival might be attributed to several factors. First, the patient received early treatment with drugs such as warfarin and an angiotensin-converting enzyme inhibitor and showed no evidence of thrombi in the pulmonary arteries, which can excessively increase pulmonary artery pressure. Second, rich collateral vessels surrounded the alveoli (Figure 5) might have decreased the pulmonary artery pressure, acting like pulmonary veins in a healthy lung. Third, the dilated pulmonary artery might have contributed to slower disease progression, as reported previously.\(^4\)

**Conclusion**

We described a long-term survivor with PVOD who had a giant pulmonary artery aneurysm. Early medical intervention may improve outcomes in patients with PVOD.

**Disclosures**

None.

**References**


![Figure 2](image-url) **Figure 2.** A computed tomographic scan showing an enlarged pulmonary artery, gradually compressing both bronchi. **A**, The scan taken in 1992. **B**, The scan taken in 2001. **C**, The scan taken in 2009.

![Figure 3](image-url) **Figure 3.** Bronchoscopy during mechanical ventilation shows the respiratory tract well opened with high positive end-expiratory pressure (PEEP; top). On the other hand, without positive airway pressure, the left and right main stem bronchi were almost completely compressed (bottom).
Figure 4. Autopsy lung revealed dilatation of pulmonary arteries (A and B; arrows). Alveolar walls were thickened with increased capillaries and hemosiderosis (C). In the interlobular septa, capillaries increased instead of pulmonary veins (C and D; arrows). BR indicates bronchus; PA, pulmonary artery; HE, hematoxylin and eosin stain; and EVG, elastica van Gieson stain.

Figure 5. Enlarged bronchial veins served as collateral pathways (A and B). BR indicates bronchus; BV, bronchial vein; HE, hematoxylin and eosin stain; and EVG, elastica van Gieson stain.
Figure 6. An axial computed tomographic scan showing ground-glass opacities, smooth thickened intralobular septa, and enlarged pulmonary veins (arrows).
Long-Term Survivor With Pulmonary Veno-Occlusive Disease
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