Proximal Pulmonary Artery Aneurysms in Patients with Pulmonary Artery Hypertension: Complicated Cases

Masahito Sakuma, Jun Demachi, Jun Suzuki, Jun Nawata, Tohru Takahashi and Kunio Shirato

Abstract

Cases with proximal pulmonary artery aneurysm (PAA) sometimes have severe complications. We report 4 cases of proximal PAA complicated by pulmonary hypertension. Three cases had proximal PAA and one had both proximal and peripheral PAA. Complications associated with proximal PAA are compression of the bronchus, dissection and/or rupture of the pulmonary artery, and thrombus of the pulmonary artery. The available medical treatments have limitations. Two of our patients with proximal PAA are awaiting lung transplantation.

Key words: dissection, thrombosis, hemoptysis

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Pulmonary artery aneurysm (PAA) is a rare disease (1). Proximal PAA occasionally induces some serious complications, including compression of the bronchus, dissection and/or rupture of the pulmonary artery, and thrombus of the pulmonary artery. Recently, the prognosis of patients with PH has improved with the development of new drugs (2-4). Moreover, as non-invasive image techniques become more widely spread, complications of PAA, which had not been recognized as big problems previously, may be clinical problems in the near future. The present report presents three cases with proximal PAA and one with both proximal and peripheral PAA.

Case Report

The first case, an 18-year-old-boy, was admitted to a local hospital at the age of 11 because of persistent cough. A huge pulmonary artery was observed on plain chest radiography, and he was diagnosed as idiopathic pulmonary artery hypertension (IPAH). He was treated with beraprost, calcium channel blocker, warfarin, and nocturnal home oxygen. At the age of 16 years, he was introduced to our institution and beraprost was replaced with epoprostenol infusion. He was classified as New York Heart Association (NYHA) functional class II. His serum level of brain natriuretic peptide (BNP) was 13.1 pg/mL (normal upper limit is 18.4 pg/mL). Chest roentgenogram showed a huge pulmonary artery Fig. 1A. Plain computed tomography revealed the compression of PAA on bilateral main bronchi Fig. 1B, and enhanced computed tomography revealed large amounts of mural thrombus in the pulmonary arteries Fig. 1C. No mutation was found in bone morphogenetic protein type II receptor gene or activin-like kinase 1 gene. At present, he is 18 years old, and awaits bilateral lung transplantation. Warfarin dose was controlled well, but his serum level of D-dimer has been consistently high (latest value was 16,800 μg/L).

The second case, a 71-year-old woman, had been diagnosed as IPAH at the age of 48 years and had been treated with warfarin, digitalis, nitrates, diuretics, calcium channel blocker, and home oxygen therapy. At the age of 71 years, her shortness of breath and leg edema worsened and she was admitted to our institute. Her serum level of BNP was 670 pg/mL. After the dose of diuretics was increased and beraprost was added, right-sided heart failure improved and her serum level of BNP was lowered to 170 pg/mL. Enhanced computed tomography on the fourth hospital day
Figure 1. Chest roentgenogram (A) and computed tomography (B and C) in Case 1. A: Note the large pulmonary artery. B: Proximal pulmonary artery aneurysm compresses the right bronchus (arrowheads). C: Thrombi are seen in the pulmonary arteries (arrows).

Figure 2. Chest roentgenogram and computed tomography in Case 2. A: Thrombus is seen in the pulmonary artery (arrow). B: Pneumonia shadow is recognized. C: Pulmonary artery aneurysm compresses the bronchus and occlusive pneumonia occurs.

showed that the inner diameter of the main pulmonary artery trunk was 80 mm and that of the right main pulmonary artery was 58 mm. PAA compressed the bilateral main bronchi and mural thrombus was recognized at the right pulmonary artery (Fig. 2A). From the 31st hospital day, she suffered from mild fever and productive cough. Chest radiography, computed tomography, and bronchoscopy revealed occlusive pneumonia due to compression of bilateral main bronchi by PAA (Fig. 2B and C). She died 15 days later.

The third case, a 17-year-old girl, was diagnosed as Eisenmenger syndrome due to atrial septal defect. At 13 years old, she was introduced to our institute and was treated with warfarin, beraprost, and nocturnal home oxygen. She was classified as NYHA functional class II. Her serum level of BNP was 23.3 pg/mL. From the age of 15, she had bloody sputum, and from 16, she suffered from severe coughing spells. Chest roentgenogram showed proximal PAA and peripheral PAA at the left lower lobe (Fig. 3A). Enhanced computed tomography (Fig. 3B) and magnetic resonance imaging showed mural thrombus in the pulmonary artery and compression of the bilateral main bronchi by PAA. Peripheral PAs appearing like a string of beads were observed in the left lower segmental artery. Three months later, she had hemoptysis and warfarin was stopped. She is now 17, and awaits bilateral lung transplantation. Her latest serum level of D-dimer was 2,900 μg/L. She has had no episode of pneumonia or sepsis throughout her clinical course.

The fourth case, a 35-year-old woman, was diagnosed as Eisenmenger syndrome due to the ventricular septal defect (catheter data had been lost) and had been treated with diu-
Figure 3. Chest roentgenogram (A), and computed tomography (B) in Case 3. A: Proximal pulmonary artery aneurysm and peripheral pulmonary artery aneurysms in the left lower lobe (arrows). B: Thrombi are seen in pulmonary arteries.

Figure 4. Macroscopic dissection and thrombus in the pulmonary artery (PA) from Case 4.

Discussion

Aneurysms in the pulmonary artery trunk, or in the bilateral main pulmonary artery are considered as proximal PAA. The present cases 1 to 4 were of this category although case 3 also had peripheral PAA. A diameter of over 4 cm was defined as PAA in the pulmonary artery trunk (5). PAA is occasionally accompanied by mural thrombus in PAA, compression on bronchus, and dissection and/or rupture of PAA. As shown in Table 1, our cases also had these complications.

An autopsy report in 1947 showed that proximal PAA was found in only eight of 109,571 cases (0.0073%) (1). However, since no prospective studies have been focused on PAA, the precise incidence is unknown. Etiologies for PAA are thought to be infection, pulmonary hypertension, medial degeneration in pulmonary artery, and arteriosclerosis (6, 7). Pulmonary hypertension is found in 66% of proximal PAA patients (8). In the past, syphilis was the main cause of infection for PAA. Pathologically, most structural vascular abnormalities are acquired degenerative diseases including medionecrosis and atherosclerosis (7). These abnormalities result from continual process of damage to and repair of the vascular structures (7). In the present Case 4, atherosclerosis...
Table 1. Catheter Data

<table>
<thead>
<tr>
<th>Clinical Diagnosis</th>
<th>Age (y.o.)</th>
<th>PCWP (mmHg)</th>
<th>mean PAP (mmHg)</th>
<th>RAP (mmHg)</th>
<th>mean AoP (mmHg)</th>
<th>CI (L/min/m²)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 IPAH</td>
<td>16</td>
<td>NA</td>
<td>60</td>
<td>8</td>
<td>82</td>
<td>2.63</td>
</tr>
<tr>
<td>Case 2 IPAH</td>
<td>50</td>
<td>4</td>
<td>60</td>
<td>88</td>
<td>88</td>
<td>3.11</td>
</tr>
<tr>
<td>Case 3 Eisenmenger Syndrome, ASD</td>
<td>17</td>
<td>2</td>
<td>95</td>
<td>8</td>
<td>72</td>
<td>1.99</td>
</tr>
<tr>
<td>Case 4 Eisenmenger Syndrome, VSD</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

Data in case 1 were measured before the use of epoprostenol. IPAH, idiopathic pulmonary artery hypertension; ASD, atrial septal defect; VSD, ventricular septal defect; PCWP, pulmonary capillary wedge pressure; PAP, pulmonary artery pressure; RAP, right atrial pressure; AoP, aortic pressure; CI, cardiac index; and NA, not available. *1 PCWP was not measured due to the existence of pulmonary thrombus. *2 Left atrial pressure was substituted for PCWP due to the existence of pulmonary thrombus. *3 Pulmonary blood flow was shown.

Table 2. Complication Associated with PAA

<table>
<thead>
<tr>
<th>Location of PAA</th>
<th>Stenosis of Bronchus</th>
<th>Dry Cough</th>
<th>Pulmonary Thrombus</th>
<th>Dissection of PAA</th>
<th>Hemothysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 Proximal</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Case 2 Proximal</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Case 3 Proximal and Peripheral</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Case 4 Proximal</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

PAA, pulmonary artery aneurysm

of pulmonary arteries was found.

Proximal PAA itself produces no symptoms unless there are complications such as compression on bronchus, dissection/rupture, or thrombus. PAA has been diagnosed by chest radiography, computed tomography, magnetic resonance imaging, and angiography. Generally, the only therapy for PAA with pulmonary hypertension is treatment for pulmonary hypertension, if there are no complications associated with PAA. On the other hand, proximal PAA is a potentially fatal condition. Wekerle et al reported a case with surgical repair of the PAA at lung transplantation (9). Medical treatments are limited in complicated cases. Two of the present cases (Cases 2 and 4) died. The remaining two cases (Cases 1 and 3) are awaiting lung transplantation.

Dissection

Dissection of proximal PAA is a life-threatening complication. Only 19% of dissections developed in patients without pulmonary hypertension (10), and about 80% occurred in the main pulmonary trunk (11). Sixty-seven cases with dissection of the proximal PAA were reported including the present case number 4 (10-13). Only ten cases (15%) were diagnosed while living. Seven of these ten are alive (three cases were surgically repaired and thromboendoarterectomy was performed in one case), one died, and the prognosis was not reported in two cases. Autopsy was performed in 52 cases, and the cause of death was cardiac tamponade in 35 cases (67%). Underlying diseases were cardiac in 36 cases, idiopathic pulmonary artery hypertension in 10, chronic obstructive pulmonary disease in 5, pulmonary thromboembolism in 4, iatrogenic (catheter-induced) in 2, idiopathic in 3, inflammation in 3, and unspecific in 4. Cardiac disease was congenital in 27 cases, and persistent ductus arteriosus was the most common. Main symptoms of dissection of proximal PAA were chest pain and dyspnea. Dissection of proximal PAA can be clinically diagnosed by computed tomography, magnetic resonance imaging, trans-thoracic echocardiography, and pulmonary angiography.

Airway compression

Dilated pulmonary artery causes airway compression. For example, large left-to-right shunts can compress mainly four sites; left lateral trachea, superior aspect of the left main bronchus, the origin of the left upper lobe bronchus, and the junction of the right intermediate and right middle lobe bronchi (14, 15). Clinically, airway compression may induce cough and dyspnea. Airway compression was recognized in three of our cases.

Thrombus in the pulmonary arteries

The relation between IPAH and pulmonary thrombi has been discussed (16). Warfarin improves prognosis in IPAH patients (17), which suggests the relation. The occurrence of thrombus in pulmonary aneurysm was also reported (18). Blood velocity is reduced in pulmonary hypertension because of low output, and blood congests in aneurysm. These changes and endothelial dysfunction (19, 20) may induce a prothrombotic state. The level of D-dimer elevated persistently in two of our cases. The elevation seems to be due to continuous formation of thrombus in the pulmonary artery.

Conclusion

Four complicated PAA patients with pulmonary hypertension were presented. Complications associated with proximal PAA were compression of bronchus, dissection and/or rup-
ture, and thrombus of pulmonary artery. To date, the available medical treatments are limited. Two of the present pa-
tients with proximal PAA are awaiting lung transplantation.

References


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