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Lung: Case Report

Reoperation of Pulmonary Artery Intimal Sarcoma for Recurrence After Pulmonary Artery Replacement

Kentaro Miyazaki, MD,[1]
Isao Matsumoto, MD, PhD,[1]
Satoshi Nishikawa, MD,[1]
Takashi Wada, MD,[1]
Tetsuya Takayama, MD,[1]
Daisuke Saito, MD, PhD,[1]
Shuhei Yoshida, MD, PhD,[1]
Kenji Iino, MD, PhD,[2] and
Hirofumi Takemura, MD, PhD[2]

Pulmonary artery intimal sarcoma has very poor prognosis, for which radical resection may be difficult. A 68-year-old man with pulmonary artery intimal sarcoma underwent bilateral pulmonary artery resection and replacement. At 31 months postoperatively, recurrence occurred at the anastomotic site of the right pulmonary artery. Therefore, he underwent right upper lobectomy and combined resection and reconstruction of the right pulmonary artery. The tumor was removed, while preserving the lungs as much as possible. He is still alive 42 months after the initial surgery.

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Pulmonary artery intimal sarcoma has a poor prognosis. Surgery is the most effective treatment. However, depending on localization of tumor, radical resection may require extensive pulmonary resection or surgery itself may be difficult. Herein, we report a case of pulmonary artery intimal sarcoma that was successfully treated with resection and reconstruction of the bilateral pulmonary arteries and repeat resection and reconstruction of the pulmonary arteries for recurrence. The tumor was removed while preserving the lungs as much as possible, resulting in long-term survival.

The patient was a 68-year-old man. He had an annual imaging test at a complete medical checkup. He had no symptoms. During complete medical checkup, positron emission tomography-computed tomography revealed fluorodeoxyglucose accumulation only in the left pulmonary artery (Figure 1A). Contrast-enhanced computed tomography (CT) revealed a low-absorption area in the same site (Figure 1B). Malignant neoplastic lesions of vascular origin, such as pulmonary artery intimal sarcoma, were suspected. A biopsy indicated a risk of damage to the pulmonary artery and concomitant pulmonary embolism. Surgery was considered necessary for a definitive diagnosis.

The surgery was performed under general anesthesia and differential lung ventilation (Figure 2A). A median sternal incision was made and cardiopulmonary bypass (CPB) was initiated. The lesion extended from the main pulmonary artery to the front of the superior trunk artery on the right side, and the A3 and A1+2 branches on the left side. After an additional left third intercostal thoracotomy, ligation, and transection of the A3 and A1+2 branches of the left pulmonary artery, the pulmonary trunk was dissected, and a vascular prosthesis (Gore-Tex 16 mm; W.L. Gore & Assoc.) was anastomosed to the peripheral side. The right pulmonary artery was transected on the central side, where the superior trunk artery branched off, and a vascular prosthesis (Gore-Tex 16mm) was anastomosed to the peripheral side. The central pulmonary artery was transected above the pulmonary valve. The vascular prosthesis was anastomosed in a Y-shape and reconstructed; the patient was weaned from CPB, and surgery was completed. The operation time and CPB time were 571 and 94 minutes, respectively. No circulatory arrest was performed.

The postoperative pathologic diagnosis was pulmonary artery intimal sarcoma (Figure 2B). There were no pathologic malignant findings in the surgical margins. The patient was discharged on postoperative day 35.

At 31 months postoperatively, positron emission tomography-computed tomography showed fluorodeoxyglucose accumulation near the anastomotic site between the vascular prosthesis and the right pulmonary artery. Contrast-enhanced computed tomography also revealed a poorly contrast-enhanced

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[1]Department of Thoracic Surgery, Kanazawa University, Kanazawa, Japan; and [2]Department of Cardiovascular Surgery, Kanazawa University, Kanazawa, Japan

Address correspondence to Dr Matsumoto, Department of Thoracic Surgery, Kanazawa University, 13-1, Takara-machi Kanazawa City 920-8641, Japan; email: isa-mat@med.kanazawa-u.ac.jp.

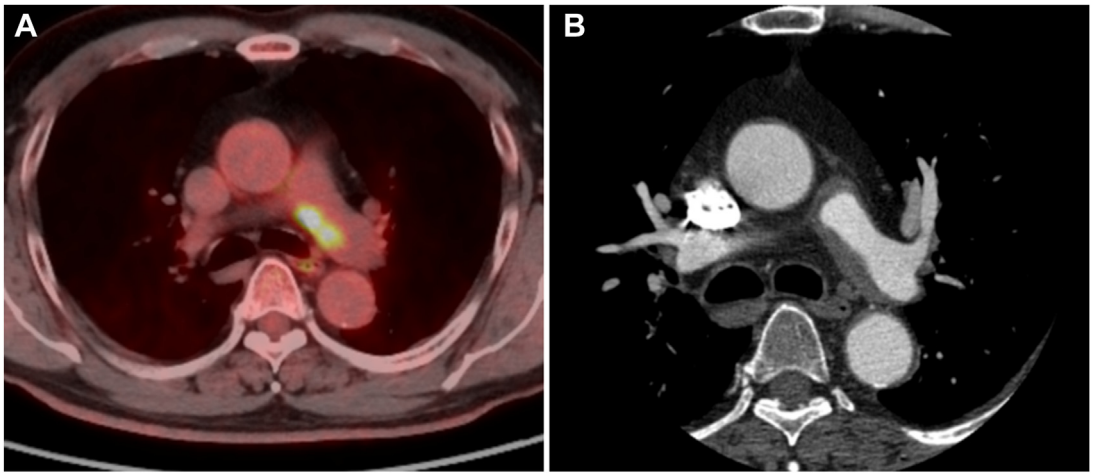

FIGURE 1 (A) Positron emission tomography-computed tomography reveals fluorodeoxyglucose accumulation in the left pulmonary artery. (B) Contrast-enhanced computed tomography reveals a low-absorption area in the same site.

area; a tumor recurrence was diagnosed. The patient had impaired respiratory function after the initial surgery, wherein right pneumonectomy was considered difficult. Systemic chemotherapy or radiation therapy were discussed as options at our cancer board. However, because the tumor was localized, we judged that radical resection was possible with right upper lobectomy if the pulmonary artery was reconstructed. Reoperation was initiated under general anesthesia and differential lung ventilation. A balloon was implanted through the right femoral vein into the vascular prosthesis of the right pulmonary artery for vascular occlusion. He was placed in the left lateral recumbent position, the fourth intercostal space was opened, and the right upper lobectomy was performed. During resection of the pulmonary artery, balloon blockade of the right pulmonary artery was attempted. However, because his circulation could not be maintained, CPB was initiated. When the pulmonary artery was cut open, a tumor was found in the lumen extending to the vicinity of the A4+5 bifurcation. The vascular prosthesis was dissected centrally from the anastomotic site and the peripheral part was resected to a grossly tumor-free area. The pulmonary artery was reconstructed using a vascular prosthesis (Gore-Tex 16 mm), the patient was weaned from CPB, and the surgery was completed (Figure 3). The operation time and CPB time were 743 and 82 minutes, respectively. No circulatory arrest was performed.

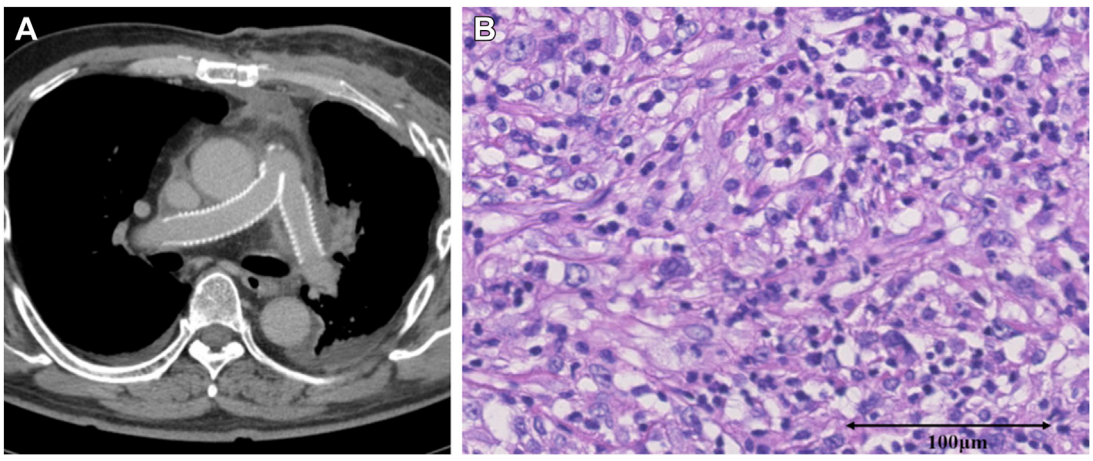

FIGURE 2 (A) Bilateral pulmonary artery resection and replacement are performed. (B) Postoperative pathology reveals pulmonary artery intimal sarcoma.

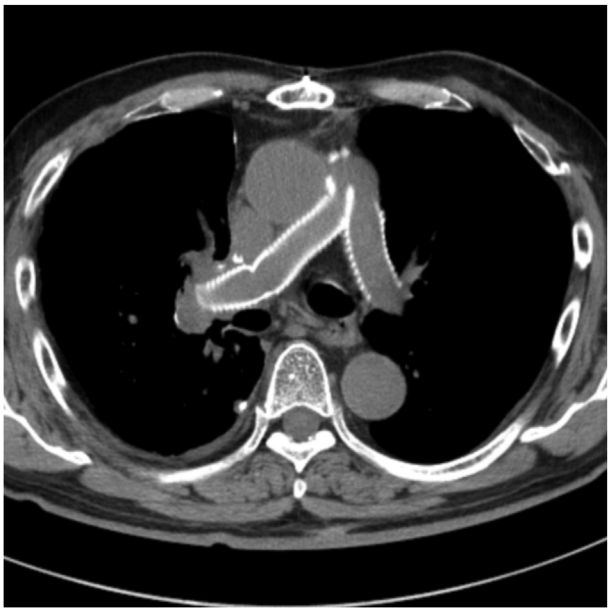

FIGURE 3 Right upper lobectomy and combined resection and reconstruction of the right pulmonary artery are performed for recurrence.

Postoperative pathologic examination revealed that the histology was similar to that of the original tumor, and the tumor was diagnosed as a recurrence of the primary pulmonary artery intimal sarcoma. There were no obvious pathologic malignant findings in the surgical margins. One month after surgery, his respiratory function declined (forced expiratory volume in 1 second: 59.6%, and vital capacity: 79.4%) and pulmonary artery pressure increased (systolic pulmonary artery pressure (PAP)/diastolic PAP/mean PAP : 44/7/21 mm Hg). Therefore, home oxygen therapy was introduced; he was discharged on postoperative day 38. Ten months after reoperation, he is currently alive without recurrence.

COMMENT

Pulmonary artery intimal sarcoma is a rare disease with a poor prognosis, with an incidence of 0.001% to 0.03%.[1] Although contrast-enhanced computed tomography is useful for diagnosis, differentiating pulmonary artery intimal sarcoma from pulmonary thromboembolism is important because their findings are similar and a misdiagnosis can occur.[2] In our case, we suspected a malignant tumor because positron emission tomography-computed tomography revealed localized fluorodeoxyglucose accumulation, echocardiography showed no right heart overload, and blood test results showed no abnormal coagulation function.

Multidisciplinary treatments, including surgery, are effective, whereas chemotherapy or radiotherapy alone is not expected to have a therapeutic effect. Patients with pulmonary artery intimal sarcoma have a median survival of 1.5 months without surgical treatment.[3] In contrast, in resectable cases, surgical resection of the tumor could extend survival from 8 to 36 months.[4] Blackmon and associates[5] reported that complete tumor resection improved prognosis to an average of 36.5 months, with a 5-year survival rate of 49.2%.

Although the surgical margins were negative, localization of the lesion at the time of recurrence suggested that small tumor cells remained in the right pulmonary artery transects. Aggressive surgical resection of recurrent lesions has been reported to improve prognosis.[6,7] In our case, the recurrent lesion was localized near the anastomosis between the right pulmonary artery and the vascular prosthesis; surgery was performed. If the right pulmonary artery is reconstructed, it can be resected via a right upper lobectomy.

During the reoperation, the middle lobe of the right lung was left in place to preserve the patient's respiratory function. Therefore, although the resection margin was pathologically negative, it was probably insufficient. Other optional treatment may be needed because of the possibility of future local recurrence. In this study we examined a case of 2 surgeries for pulmonary artery intimal sarcoma or its recurrence to date, with a long-term survival. The extent of the lesion and patient's condition should be considered, and surgery should be performed while preserving patient's cardiopulmonary function.

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PATIENT CONSENT
Obtained.

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