

Case Report

Primary Pulmonary Artery Leiomyosarcoma : a Case of 4-year Survivor after Surgical Treatment

Hironori Ishida¹, Hideo Adachi², Shinichiro Okada¹, Seiichirou Murata²,
Takeshi Ishida³, Shigeki Yamada⁴, Fumio Konishi¹

Pulmonary artery sarcoma is a rare and usually fatal tumor. We report a case of a 54-year-old female with an intraluminal sarcoma which occluded the left pulmonary artery and narrowed the right pulmonary artery and pulmonary trunk. Under cardiopulmonary bypass, the patient underwent resection of the right pulmonary artery and pulmonary artery trunk including the tumor, pulmonary artery reconstruction, and left pneumonectomy. Final pathologic diagnosis was leiomyosarcoma. The patient survived at home for 4 years 8 months, until she died of tumor recurrence. Radical surgical resection under cardiopulmonary bypass probably improves symptoms and prolongs postoperative survival.

(Keywords : pulmonary artery sarcoma, leiomyosarcoma, pulmonary thromboembolism, surgery, survival)

I. Introduction

Sarcoma of the pulmonary artery is a rare disease, and its prognosis is reported to be poor. We encountered a case of pulmonary artery sarcoma with complete occlusion of the left main pulmonary artery and stenosis of the right pulmonary artery. Because of the rarity of this disease and the relatively long survival of the present patient after surgery, we report this case with a review of the literature.

II. Case report

The patient was a 54-year-old female whose main complaints were chest discomfort and hoarseness. The patient had undergone surgery for endometrial carcinoma at the age of 45, and she had had hypertension and hyperlipidemia since the age of 50. There were no particular illnesses in her family history, and she had no history of smoking. In October 1998, the patient began to suffer from chest discomfort. A chest X-ray taken at a local clinic showed a mediastinal mass on the left side.

The patient was referred to our hospital in December 1998, when her hoarseness appeared. In January 1999, the patient was admitted to our hospital for further examination and treatment.

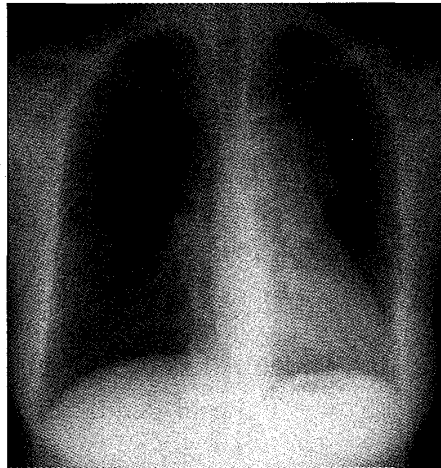


Figure 1. A chest X-ray on admission shows a mass in the region of the left hilus to the mediastinum on the left side.

At the time of admission, the patient's height was 158 cm and her weight was 57 kg. Blood pressure was 160/74 mmHg, pulse rate was 80 beats/min, and there were no signs of arrhythmia, anemia, jaundice or cyanosis. There were no palpable swollen lymph nodes. Respiratory sound was clear, but there was a systolic ejection murmur in Levine 3 degree in the second intercostal space on the left side of the sternum.

Laboratory data showed no abnormalities other than a PaO_2 level of 69.2 mmHg in arterial blood gas analysis. Results of the pulmonary function test were normal. Serum concentrations of tumor markers such as CEA, CA19-9 and CYFRA were within normal limits.

A chest X-ray on admission showed a mass in the region of the left hilus to the mediastinum on the left side. Compared to the right side, left lung volume was slightly decreased and the mediastinum was shifted to the left (Fig. 1).

A chest computed tomography (CT) showed a low-density mass filling the lumen of the pulmonary artery trunk and extending into the left and right main pulmonary arteries (Fig. 2). The portion of the mass in the left main pulmonary artery extended into the space surrounding the pulmonary artery. These findings suggested the presence of a neoplastic lesion. CT of the brain and abdomen showed no distant metastases. Bone scintigraphy showed no abnormalities.

Bronchoscopy showed paralysis of the left vocal cord, and the mucosa of the trachea and bronchi were normal.

Pulmonary arteriography showed complete occlusion of the left main pulmonary artery and severe stenosis of the right main pulmonary artery (Fig. 3). The difference in systolic pressure between the right ventricle and the pulmonary artery was 50 mmHg, which is unusually high. The diagnosis from blood aspiration cytology using an endovascular catheter was sarcoma.

Based on the results of preoperative examinations, a diagnosis of pulmonary artery sarcoma was made. To prevent sudden death due to complete occlusion of the pulmonary arteries, surgical resection was performed in February 1999.

The surgery was performed with a median sternal incision under complete cardiopulmonary bypass (Fig. 4). When the pulmonary artery was incised, it was found that the tumor extended

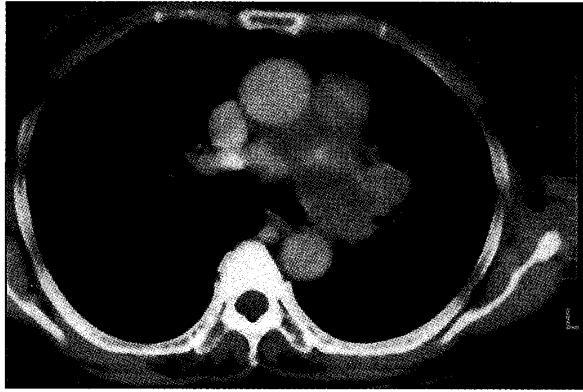


Figure 2. A chest CT scan reveals a low-density mass filling the lumen of the pulmonary artery trunk and extending into the left and the right main pulmonary arteries. The portion of the mass in the left main pulmonary artery extended into the space surrounding the pulmonary artery.



Figure 3. Pulmonary arteriography reveals complete occlusion of the left main pulmonary artery and severe stenosis of the right main pulmonary artery.

to the left semilunar valve of the pulmonary artery. The tumor arose in the pulmonary artery trunk and extended into the left and right main pulmonary arteries and into the left branches of the pulmonary artery. Because of these findings, the pulmonary artery trunk was resected including the left semilunar valve. The right main pulmonary artery was transected immediately proximal to the branching of the right upper trunk of the pulmonary artery (A^{1+3}), and a woven double velour vascular graft 22 mm in diameter was positioned between the pulmonary artery trunk and the right pulmonary artery. Following this procedure, the pericardium was incised and the left superior and inferior pulmonary veins were divided within the pericardial space. Finally, the left main bronchus was divided and the left lung was totally removed. The defect of the pericardium was repaired using a Gore-Tex pericardial sheet. A pedicled omental graft was positioned between the stump of the left main bronchus and the vessel graft.

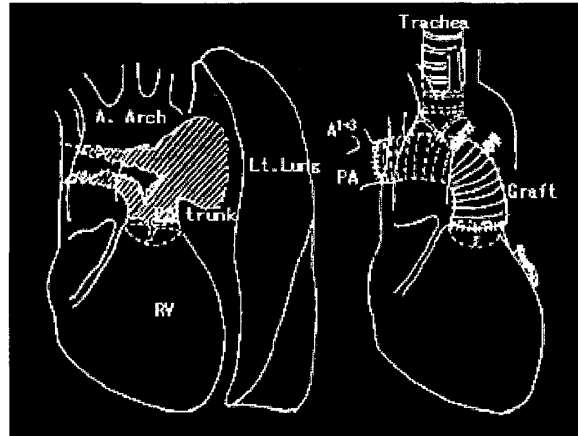


Figure 4. Anatomical schema before (left) and after (right) operation. The operative procedure was resection of the right pulmonary artery and the pulmonary artery trunk including the tumor (oblique lines), with pulmonary artery reconstruction and left pneumonectomy, under cardiopulmonary bypass.

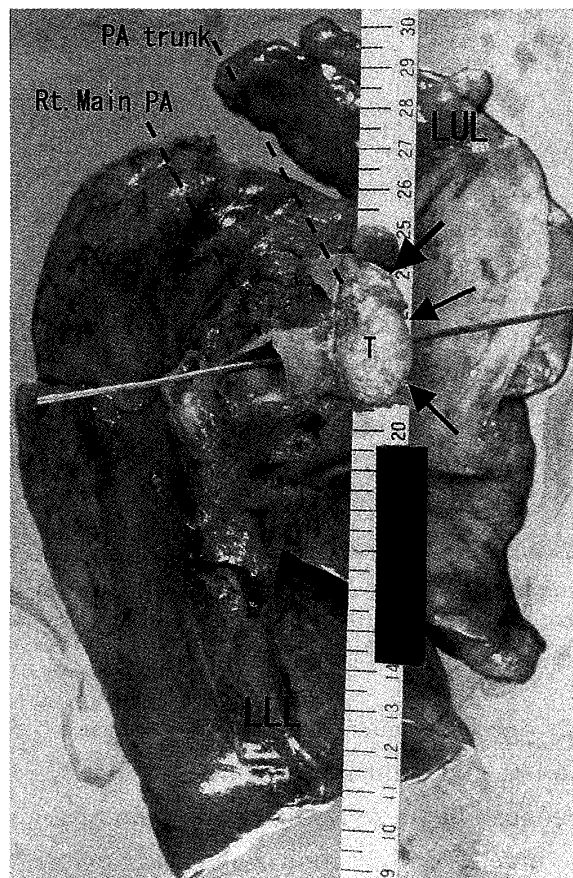


Figure 5. Resected specimen. A yellowish white polypoid tumor (arrow) was seen protruding from the lumen of the pulmonary artery on the mediastinal side of the left lung.

Figure 5 shows the resected specimen. A yellowish white polypoid tumor protruded from the lumen of the pulmonary artery at the mediastinal side of the left lung.

Figure 6 shows histological findings of the tumor. Spindle-shaped and pleomorphic atypical cells were arranged in bundles oriented in various directions. The nuclei of the tumor cells were

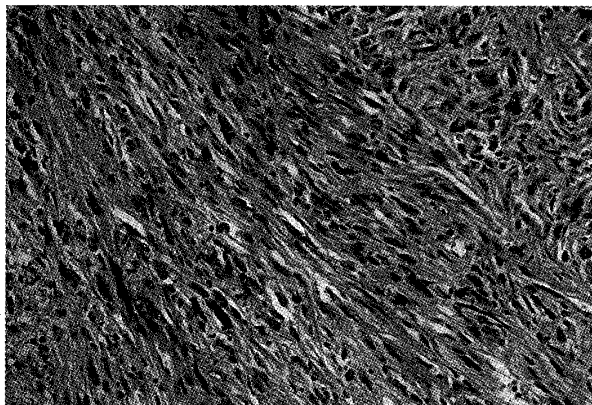


Figure 6. Histological findings. Spindle-shaped and pleomorphic atypical cells were arranged in fascicles or interlacing bundles. (H.E.×100)

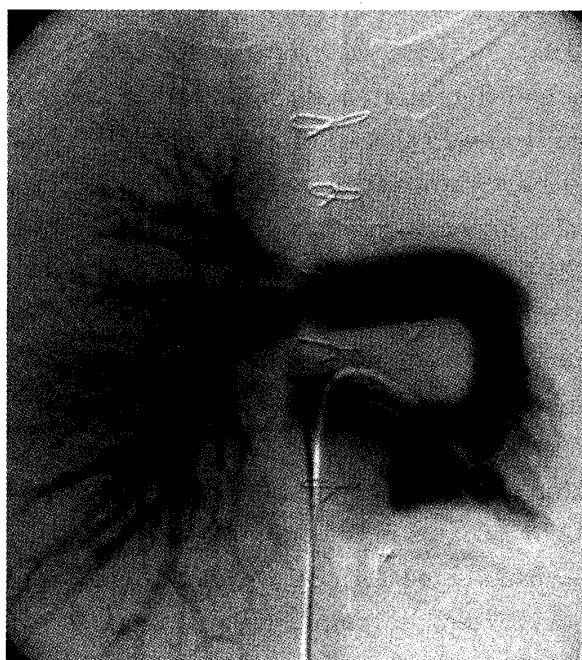


Figure 7. Postoperative pulmonary arteriography shows satisfactory blood flow within the vessel graft.

rich in chromatin, had blunt margins and had an oblong shape. In immunohistochemistry, the tumor cells stained positive for vimentin and α SMA, and stained negative for desmin, S100, pankeratin, LCA, CD34 and factor VIII. Based on these findings, a histological diagnosis of leiomyosarcoma of the pulmonary artery was made. There were the histologically negative resection margins in both peripheral and central directions

Postoperative pulmonary arteriography showed satisfactory blood flow within the vessel graft (Fig. 7). The patient was discharged 26 days after the operation. No postoperative anti-coagulation therapy or adjuvant chemotherapy was administered. In the postoperative period, the patient experienced no problems with activities of daily life other than hoarseness. Regular check-ups were performed in the outpatient clinic. Although the patient was asymptomatic, a chest CT scan performed 3 years 4 months after the operation revealed a local

recurrence of sarcoma extending from the graft into the periphery of the interlobar pulmonary artery. The patient had died of right-side heart failure 4 years 8 months after the operation without any cancer treatment.

III. Discussion

Sarcoma of the pulmonary artery is a rare disease, and only a few more than 100 cases have been reported in the literature¹⁾²⁾. Because most pulmonary artery sarcoma arises in the pulmonary artery trunk, it has been assumed that it originates in multipotential mesenchymal cells at the site of the bulbus cordis of the pulmonary artery trunk³⁾⁴⁾. The tumor is also known as "intimal sarcoma" because of its origin in the vascular intima. The tumor usually grows within the vessel lumen, and therefore should be distinguished from "mural sarcoma"⁵⁾. In a case report by Nonomura with a review of 110 cases in the literature, pulmonary artery sarcoma was found to occur slightly more frequently in females than in males (ratio, 1 : 1.3) ; the mean age at occurrence was 50 years, with a range of 21 to 81 years ; and the most frequent histological diagnosis was undifferentiated sarcoma, which accounted for one-third of the histological diagnoses of the reviewed cases, followed by leiomyosarcoma and fibrosarcoma or rhabdomyosarcoma¹⁾. Studies have shown that cases of pulmonary artery sarcoma with different histological types usually have generally similar clinical characteristics and tumor prognoses⁶⁾.

The first symptoms of this tumor are usually dyspnea, chest pain, cough and bloody sputum, which are the symptoms of pulmonary embolisms. Because the symptoms are similar to those of a chronic pulmonary artery thromboembolism, early definitive diagnosis is often difficult. In most cases, diagnosis is made by examining resected specimens or autopsy specimens¹⁾⁷⁾. In the present case, the diagnosis was made by cytology, using the blood sample taken at the time of the angiography and catheter examinations, as some authors previously reported⁸⁾⁹⁾¹⁰⁾.

Because the tumor usually originates in the pulmonary artery trunk or the pulmonary artery valve, surgical resection is usually performed with cardiopulmonary bypass. In previously reported cases of pulmonary artery sarcoma, either endarterectomy, pneumonectomy or resection of the pulmonary artery with reconstruction was performed, depending on the extent of tumor invasion and the presence or absence of pulmonary metastasis²⁾⁷⁾.

The prognosis of pulmonary artery sarcoma is extremely poor. In a review of 93 cases, Krüger⁷⁾ reported that the median survival period was 1.5 months. Among those 93 cases, there were 27 in which surgical resection was performed. The median survival rate for the patients who underwent surgical resection was 10 months longer than that of the cases without resection. Thus, it appears that the survival period can be extended by performing surgical resection, but long-term survival is rarely achieved.

The main causes of death after surgery for pulmonary artery sarcoma are local recurrence and pulmonary metastasis. Therefore, when the pulmonary artery is resected, it is important to obtain a tumor-free margin. In a report by Mayer²⁾, pneumonectomy was performed in 2 cases in which a frozen section diagnosis was positive at the resection margin, and one of the patients survived for 35 months without recurrence. In the present case, we resected as much of the right main pulmonary artery as possible, and we removed the pulmonary artery valve on

the central side. However, despite the histologically negative resection margins in both peripheral and central directions, there was local recurrence of the tumor in the present case.

There have been reports of cases of pulmonary artery sarcoma in which preoperative or postoperative chemotherapy or radiation therapy was effective²⁾⁷⁾¹¹⁾, but the effectiveness of such treatments has not been confirmed.

Long-term survival after resection of pulmonary artery sarcoma is rare. There have been only 3 reported cases in which the patient survived longer than 4 years after the operation²⁾¹²⁾¹³⁾. In those 3 cases, in which it appears that there was no recurrence, pulmonary artery resection was performed under cardiopulmonary bypass to remove the tumor, and pneumonectomy was not performed. Thus, those 3 cases suggest that complete resection is possible if the tumor is detected at an early stage.

In the present case, the tumor recurred within the pulmonary artery 3 years 4 months after the operation, and the patient died of heart failure at 4 years 8 months after the operation. However, no distant metastasis or pulmonary metastasis was detected at any time. The present results suggest that complete resection of the tumor can help prevent early sudden death and provide relatively long postoperative survival.

IV. Conclusions

Pulmonary artery sarcoma causing stenosis or occlusion of the pulmonary artery should be completely resected using cardiopulmonary bypass to provide long time survival in the selected patients.

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原発性肺動脈平滑筋肉腫：4年間の生存を得た1切除例

石田 博徳¹ 安達 秀雄² 岡田晋一郎¹ 村田聖一郎²
石田 岳史³ 山田 茂樹⁴ 小西 文雄¹

要 約

肺動脈肉腫は稀で致命的な疾患である。54歳女性で、左主肺動脈の完全閉塞と肺動脈主幹部から右肺動脈の狭窄を来す血管内肉腫の症例を報告する。体外循環下に、右室-右肺動脈の人工血管置換、左肺全摘術を行った。最終病理診断は平滑筋肉腫であった。術後4年8ヶ月で再

発死亡するまでは、自宅での日常生活が可能であった。できる限り完全切除を試みることで、早期の突然死を回避でき、比較的長期の生存が得られる可能性がある。

(キーワード：肺動脈肉腫，平滑筋肉腫，肺動脈血栓塞栓症，手術，生存)

1. 自治医科大学附属大宮医療センター 一般・消化器・呼吸器外科
2. 自治医科大学附属大宮医療センター 心臓血管外科
3. 自治医科大学附属大宮医療センター 総合診療科
4. 自治医科大学附属大宮医療センター 病理