Kobe University Repository : Kernel

PDF issue: 2026-11-02

Primary Leiomyosarcoma of the Pulmonary Artery

Inomata,Takumi ; Kawata,Masahito ; Tabuchi,Hikaru ; Sawatari,Hisayuki ; Gotoh,Takeo ; Furumoto,Masaru ; Ogawa,Kyoichi ; Maeda,Kazumi

(Citation)
Bulletin of allied medical sciences Kobe : BAMS (Kobe),4:117-123

(Issue Date)
1988-12-26

(Resource Type)
departmental bulletin paper

(Version)
Version of Record

(URL)
https://hdl.handle.net/20.500.14094/80070300

KOBE UNIVERSITY

Primary Leiomyosarcoma of the Pulmonary Artery

Takumi Inomata[1], Masahito Kawata[1], Hikaru Tabuchi[1], Hisayuki Sawatari[1], Takeo Gotoh[1], Masaru Furumoto[2], Kyoichi Ogawa[2] and Kazumi Maeda[3]

Primary Leiomyosaromas of the pulmonary artery are extremely rare and the correct antemortem diagnosis of them is seldom made. We present a case of leiomyosarocma arising from the posterior wall of the right pulmonary artery in a 35 years old male.
He was hospitalized for further examination of exertional dyspnea, chest pain and cough associated with hemoptysis of a month's duration. On admission, pulmonary thromboembolism was strongly suspected by echocardiography, lung scintigraphy, cardiac catheterization and pulmonary artery angiography. However, a tumor was found to be situated in the right pulmonary artery during the thromboectony and a histological examination revealed that it was leiomyosarcoma.
The patient was free of symptom for a while, but expired of right heart failure two months after operation.
The pulmonary sarcoma should be taken into consideration in the differential diagnosis of pulmonary embolization.

Key Words
Leiomyosarcom,
Pulmonary artery,
Pulmonary artery angiography,
Lung scintigraphy.

INTRODUCTION

Primary leiomyosarcomas of the pulmonary artery artery are rare tumors and the manifestation of the disease may mimic the common condition in patients with primary pulmonary hypertension, pulmonary valvular stensis (including pulmonary peripheral stenosis) and pulmoanry embolism (1). The originating site of this tumor has been reported to be usually main pulmonary trunk (2,3).

We describe a 35 year old male with pulmonary leiomyosarcoma originated in the right pulmonary artery and provide reviews of literature associated with it.

CASE PRESENTATION

A 35 year old male was admitted to the Takasago Municipal Hospital on November 25, 1984 with exertional dyspnea, chest pain and hemoptysis of a month's duration.

On admission, physical examination revealed that the patient was dyspneic on minimal exertion and had a regular pulse rate of 98 beats/min and a blood pressure of 100/60 mmHg. A grade of 3/6 middle pitched ejection murmur at the second to fourth intercostal space near the left sternal border was audible, the pulmonary second sound was split and accentuated. The ECG showed

Department of Internal Medicine, Takasago Municipal Hospital[1], Hyogo, Himeji Cardiovascular Center[2], Hyogo and School of Allied Medical Sciences, Kobe University[3], Kobe, Japan.

sinus rhythm and inverted T wave in leads Ⅲ and V_{1-4}, and transitional zone between lead V_4 and V_5 (Fig. 1). Chest X-rays revealed a cardiothoracic ratio of 47% and no abnormal findings in the lung field. Laboratory investigation showed the following : red blood cells 571 × 10^4, hemoglobin 17.7 g/dl, white blood cells 6900/mm^3, erythrocyte sedimentation rate 10 mm/h. C-reactive protein 1+, glutamic oxalacetic transaminase 29U (normal 8-4), glutamic pyruvic transaminase 42U (normal 5–35), lactic dehydrogenase 506U (normal 50–400), creatinine phosphokinase 34U (normal 80). Although respiratory function test showed no abnormality, the arterial blood gas showed a pH of 7.41, PaO_2 of 61 mmHg and $PaCO_2$ of 35 mmHg under room air at rest and PaO_2 declined to 45 mmHg after about 120 meters' walk.

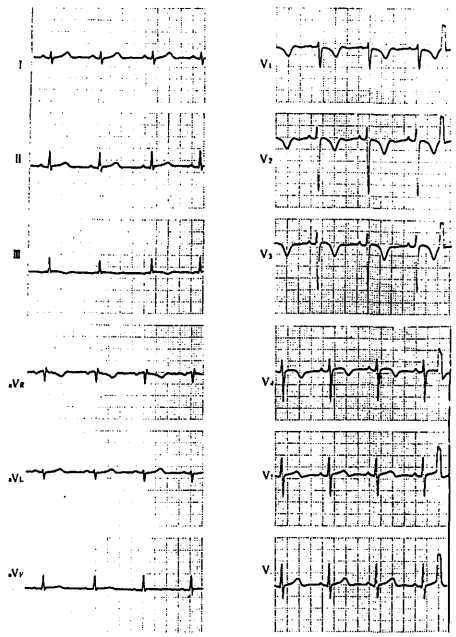


Fig. 1. An electrocardiogram on admission showed sinus rhythm at a rate of 68, mean QRS axis of 80°, S wave in lead Ⅰ, T wave inversion in leads Ⅲ and V_{1-4}, and transitional zone between lead V_4 and V_5.

B-mode and M-mode echocardiography showed slight enlargement of the right ventricle. Mild tricuspid and mild pulmonary regurgitation were confirmed by contrast echocardiography and pulse doppler echocardiography. A scintigram of the lung with ^{99m}Tc-MAA was performed and demonstrated an almost complete defect of the right main pulmonary artery, suggestive of the makedly reduced blood flow of the right upper pulmonary artery and the right lower pulmonary artery (Fig. 2 left side).

Right heart catheterization and pulmonary artery angiography were performed and a pressure of the right atrium was 9/10 mmHg, mean 4, the right ventricle 68/–4, end-diastolic pressure 8, the pulmonary artery 71/14, mean 36 and the pulmonary artery wedge pressure was 10/5, mean 7.

Cardiac index was 2.61 ℓ/min/m^2 by thermodilution method. On the clinical history and examinations, this case was strongly suspected of pulmonary thromboembolism. Therefore urokinase of 960,000U was injected selectively into the right pulmonary artery for an hour but the unopacified lesion remained unchanged (Fig. 3).

Furthermore, heparine of 20,000 U/day for four days and urokinase of 240,000–360,000 U/day for ten days were administered intravenously, but his symptons did not improve at all.

Abdominal echocardiography, abdominal computed tomography, esophaggiogastric fluoroscopy and venous angiography of inferior vena cava and

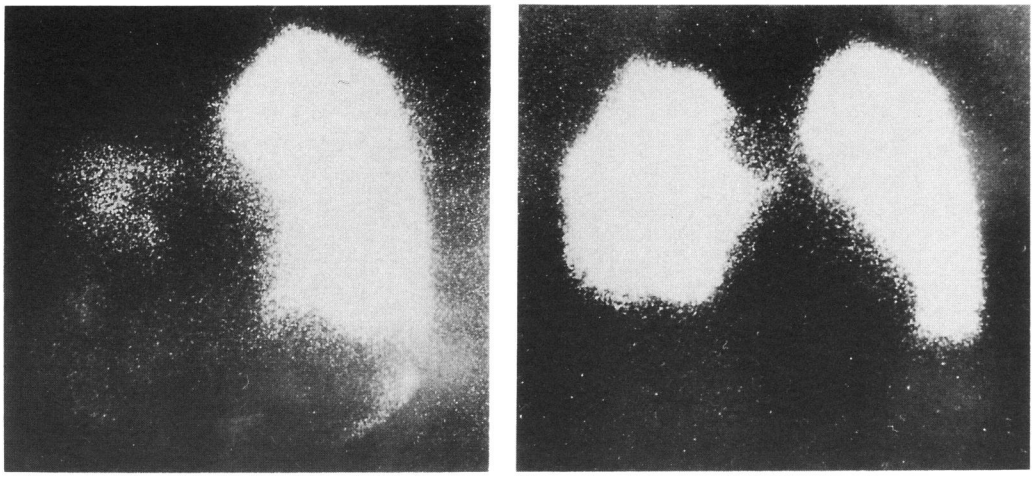


Fig. 2. Radionuclide scintigram of the lung with ^{99m}Tc-MAA before operation demonstrated an almost complete defect of the right main pulmonary artery (left). Radionuclide scintigram four days after the operation showed remarkable increase in the blood flow in the right lung except for that part of the right upper lobe (right).

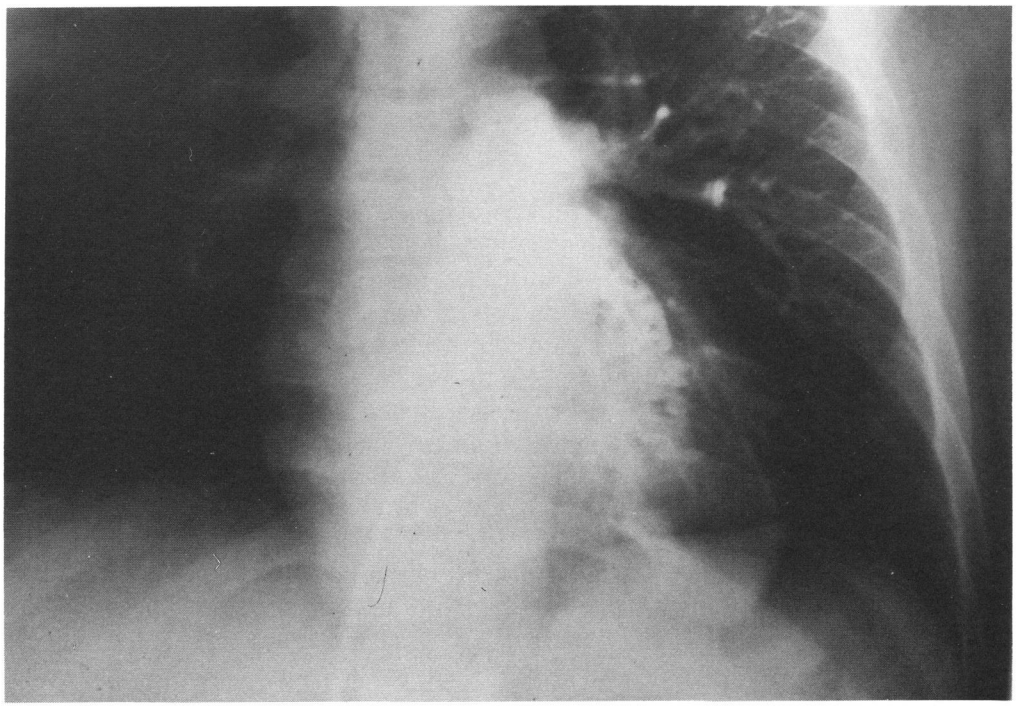


Fig. 3. Pulmonary angiography showed an almost complete defect of right main pulmonary artery.

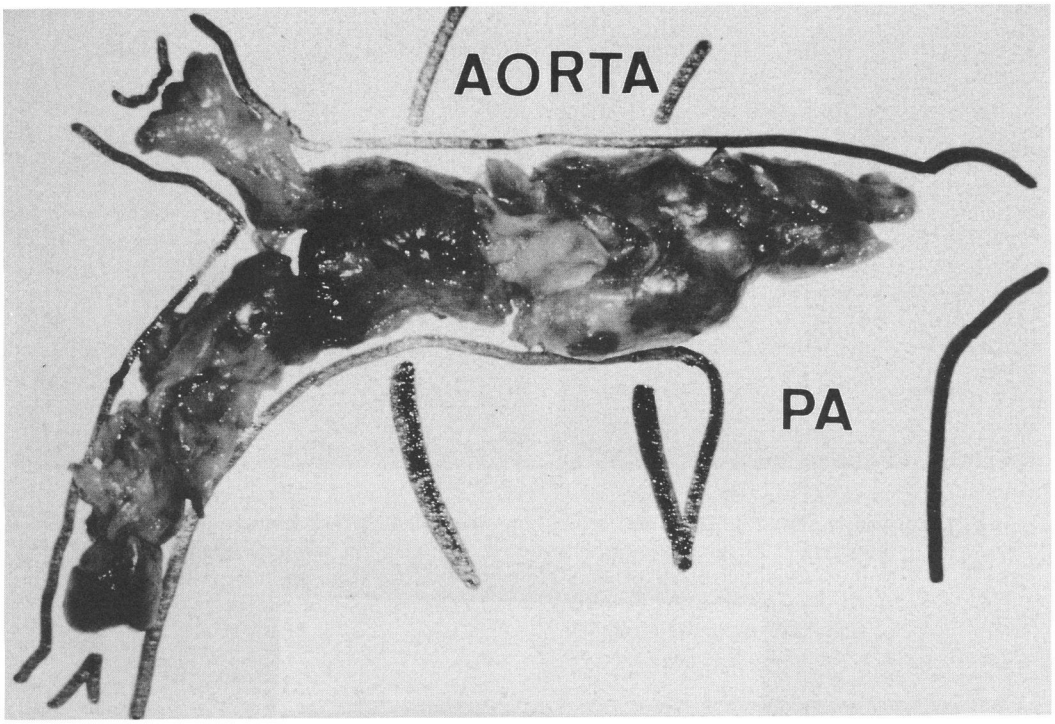


Fig. 4. Yellow smooth-surfaced tumor-like masses in the right pulmonary artery were removed except for that in the right upper pulmonary artery.

bilateral lower extremities showed no abnormal findings, thereby suggesting of thromboembolism of unknown origin.

Thromboembolectomy under extracorporeal circulation was performed on December 20, 1984, at the operation yellow smoothsurfaced tumor-like masses in the right pulmonary artery were removed except for that in the right upper pulmonary artery (Fig. 4).

The light microscopic findings showed tumor cells with eosinophilic spindle-shaped cytoplasm arranged fascicularly and partially displaying the cart-wheel pattern of tumor cells. The nuclei were rugby football-like or spindle shaped, and had mild pleomorphism and mitoses. Each tumor cell was surrounded with fine reticulum fibers.

Many cells contained PAS positive granules (Fig. 5). On the basis of the above histologycal findings, the tumor in the right pulmonary artery was confirmed as leiomyosarcoma.

After the operation, the pulmonary artery pressure was 46/13, mean 25 mmHg and the cardiac index was 3.3 ℓ/min/m^2. Subjectively, the patient's dyspnea was relieved. Radionuclide scintigram of the lung four days after the operation showed remarkable increase in the blood flow in the right lung except for the in a part of the right upper lobe (Fig. 2 right side).

10 mg/day of adriamycin was administrated for five days after the operation. However, a temperature of 37–38°C continued after the operation,

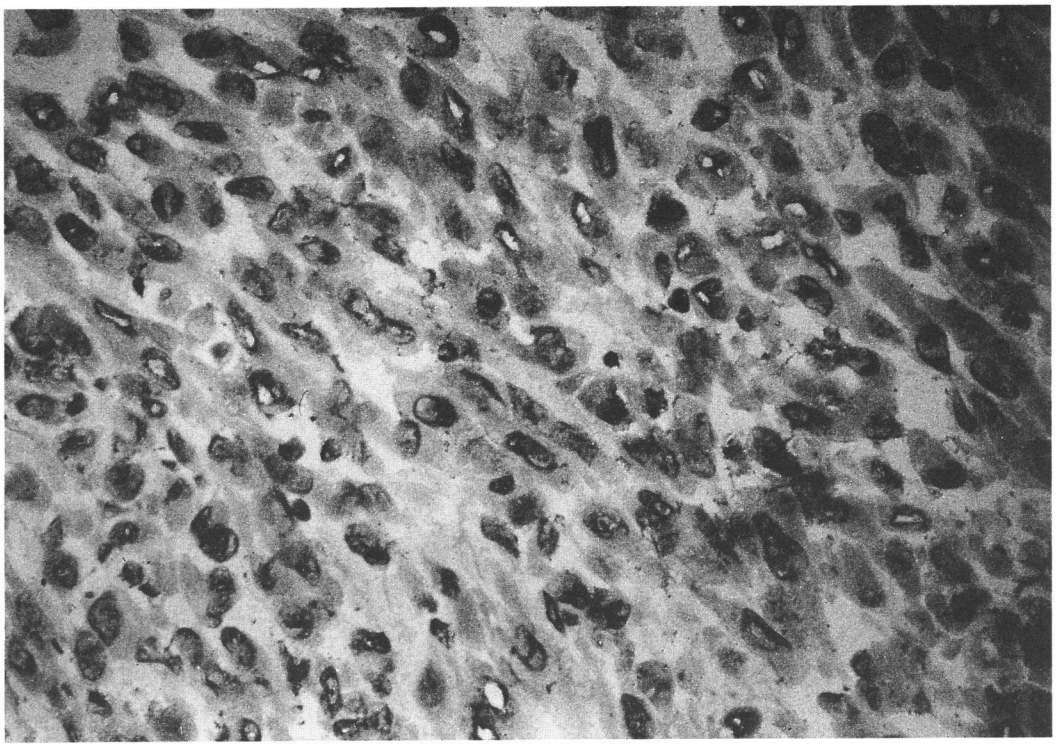


Fig. 5. The light microscopic findings showed tumor cells with eosinophilic spindle-shaped cytoplasm arranged fasciculary. ×200

therefore antibiotics and febrifuges were administered. Whole body radionuclide scintigram with Ga revealed no abnomal findings.

On Jan. 25, 1985, pulmonary artery angiogram revealed a partial defect of the right main pulmonary artery and total obstruction of the right upper pulmonary artery, which suggested a relapse of the tumor. Pulmonary artery pressure was 59/15, mean 33 mmHg.

On Feb. 22, 1985 (about two months after the operation) the patient vomitted after minimal exercise. His systolic blood pressure fell to 80 mmHg, and he felt into shock state. On pulmonary artery angiogram on Feb. 23, 1985, the pulmonary trunk only was opacified. Several hours after the angiography he died.

Autopsy was carried out two hours after his death. The tumor (2×2×8cm) originated in the posterior wall of the right main pulmonary artery and invaded intraluminally to both right and left ventricular muscle. No tumor was observed in any other. Histolgical diagnosis was the same as that of the operation.

DISCUSSION

Primary pulmonary artery leiomyosarcoma is difficult to diagnose before autopsy. And 35 cases of primary pulmon-



Fig. 6. The tumor origianted in the posterior wall of the right main pulmonary artery (arrows).

ary artery sarcoma were reported in the world by 1984. In 1984 Pain et al (4) reviewed 19 cases of leiomyosarcoma and angiographic studies were performed in 5 cases of them. Only one case was diagnosed preoperatively. All cases had tumors originated in the pulmonary artery trunk with one exception originating in the pulmonary artery. The present case was suspected of pulmonary thromboembolism on admission. However, the chest X-ray did not show any abnormality. Right ventricular loading and pulmonary hypertension based on echocardiography and right heart catheterization were suggesting of pulmonary stenosis (including pulmonary peripherol stenosis) primary pulmonary hypertension, pulmonary thromboembolism, however, the findings of echocardiography and pulmonary artery angiography leaded to the most likely diagnosis of pulmonary embolism.

Embolectomy was performed. Histological study showed fascicular and partial cart-wheel pattern of spindle-shaped tumor cells, and the nuclei displaying plemorphism and mitosis. On autopsy, the tumor invading intraluminally from the posterior wall of the right main pulmonary artery was defined as primary, Also, we observed several metastases, which were not found during the operation.

The prognosis of primary pulmonary artery sarcoma is poor and chemotherapy and radiation have no effect. All cases reported previously died in three

years after the beginning of symptoms. In this case, operation and chemotherapy were performed but a relapse was found about a month after the operation. It is reported that primary pulmonary artery sarcoma contains fibrosarcoma, osteosarcoma, rhabdomyosarcoma, myxosarcoma, angiosarcoma, etc (5). In every case, the preoperative diagnosis is very difficult. Reports of a diagnosed case before autopsy are rare (6). Primary pulmonary artery sarcoma should be considered as one of the differential diagnoses of pulmonary thromboembolism.

Moreover, it is important to diagnose as early as possible using echocardiography, radionuclide scintigraphy and angiography.

REFERENCES

1. Hynes Jk, Smith HC, Holmes DR, et al : Preoperative angiographic diagnosis of primary sarcoma of the pulmonary artery. Circulation 66, 672-674, 1982
2. Hayata T, Sato E : Primary leiomyosarcoma arising in the trunk of pulmonary artery. Acta Path Jpn 27, 137-144, 1977
3. Iwasaki I, Iwase H, Horie H, et al : Leiomyosarcoma of pulmonary truncus. Acta Patho Jpn 34, 863-867, 1984
4. Pain J, Sayer RE : Primary leiomyosarcoma of the pulmonary artery. Eur J Respir Dis 65, 139-143, 1984
5. McConell TH : Bony and cartilaginous tumors of the heart and great vessels. Cancer 25, 6611-617, 1970
6. Wright EC, Wellons HA, Martin RP : Primary pulmonary artery sarcoma diagnosed noninvasively by two-dimensional echocardiography. Circulation 67, 459-462, 1983