

## Primary Pulmonary Artery Sarcoma Detected with a Pulmonary Infarction

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### Abstract

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Primary malignant tumors of the pulmonary arteries occur infrequently. The clinical presentation and diagnostic imaging features of the tumor are usually nonspecific and correct diagnosis is often delayed. In this report, we present a case of pulmonary artery sarcoma. MRI and PET-CT were found to be useful for differentiating the tumor from a thromboembolism.

**Key words:** pulmonary artery sarcoma, PET-CT, MRI

(DOI: [10.2169/internalmedicine.46.6292](https://doi.org/10.2169/internalmedicine.46.6292))

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### Introduction

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Primary sarcomas of the pulmonary artery are uncommon and remain an interesting subject of case reports. Because of the rare incidence and nonspecific clinical manifestations, pulmonary artery sarcoma (PAS) is often mistaken for other diseases, the most frequent of which is a pulmonary embolism. In this case, we diagnosed PAS based on both the results of a coagulation survey and its clinical course with the help of some useful radiological modalities.

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### Case Report

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A 53-year-old woman was admitted for an abnormal shadow on a chest X-ray, dyspnea on exertion, and a one-time experience of bloody sputum. Two months prior to admission, a non productive cough and dyspnea on effort developed. Prior to that time she had been in good health. After one week, she had a 38.0°C fever for two days and visited a local hospital. Chest X-ray showed no abnormal shadows, and she was treated for a common cold. One month before admission, a lancinating chest pain on the back of the right chest developed, however, the symptom resolved spon-

taneously within a week. A week before admission, non-productive cough and bilateral chest pain developed, and bloody sputum once developed. She visited the local hospital again, and a chest X-ray this time revealed a sub pleural round consolidation in the right lower lung field (Fig. 1). She was referred to our hospital to undergo a complete medical examination.

On physical examination, the patient showed a normal respiratory rate (20 breaths per minute), blood pressure of 125/78 mmHg, heart rate 75 beats per minute, body temperature of 37.3°C. Cardiac auscultation revealed a Levine II to III/IV systolic murmur over the left sternal border. An electrocardiogram showed V1 R/S above 1.0 and clockwise rotation pattern, but did not show a S<sub>1</sub>Q<sub>3</sub>T<sub>3</sub> pattern or ST-T abnormality. An arterial blood gas analysis while the patient was breathing room air in sitting position showed a partial pressure of oxygen of 69.7 mmHg. The round consolidation seen in the right lower lung field was enlarged in comparison to that before admission.

We performed a contrast-enhanced spiral computed tomographic (CT) scan, showing massive filling defects in the right pulmonary artery, with a patent left pulmonary artery (Fig. 2). Wedge-shaped subpleural lesions were found only in the right lung field. A ventilation-perfusion scintigram

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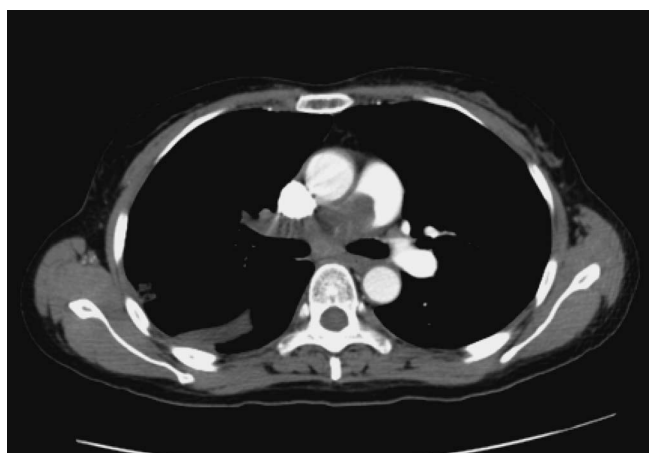
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Received for publication October 6, 2006; Accepted for publication January 17, 2007

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**Figure 1.** A roentgenogram of the chest when nonproductive cough, chest pain on both chest sides and hemoptum developed, showing a round consolidation in the lower lung field, reduced radiolucency in the middle lung field on the right side, and prominence of the right pulmonary artery.



**Figure 2.** Chest spiral CT on admission showing massive filling defects over the right pulmonary trunk and proximal arteries.

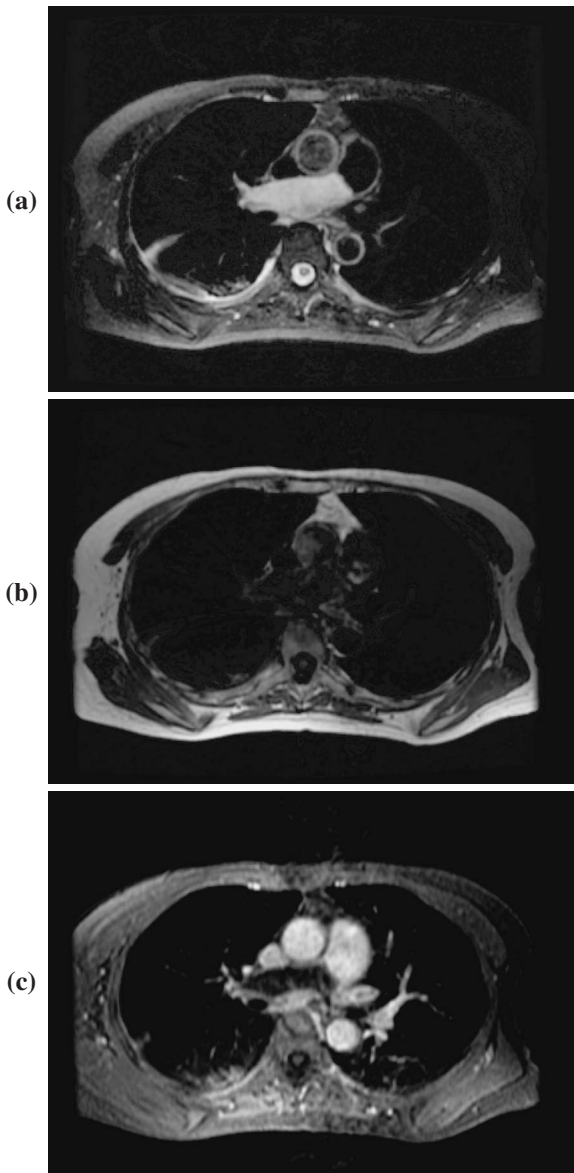
showed a reduced perfusion of the right lung field and wedge-shaped defects of ventilation. We initially suspected a pulmonary infarction due to thromboembolism. However, echocardiography demonstrated a non-dilated right atrium and right ventricle. A venous duplex examination for deep vein thrombosis was negative. The findings of surveys for coagulopathy, including antinuclear antibody and anticardiolipin antibody, are shown in Table 1. The FDP (Fibrin degradation product)-D-dimer level was slightly elevated, however, it improved to within the normal range 12 h after admission, which suggested that a thromboembolism was not strongly suspected because normal D-dimer, which would rule out an acute pulmonary embolism, is defined as under  $\leq 0.5 \mu\text{g/ml}$  (1, 2).

We performed chest MRI, which has been suggested to be

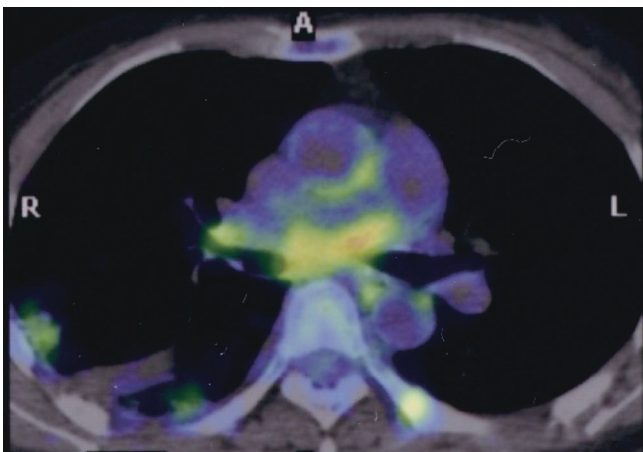
**Table 1.** Coagulation Survey on Admission

APTT	35.8 s
PT	12.2 s
Fibrinogen	448 mg/dl
FDP	5.2 $\mu\text{g/ml}$
FDP-DD	2.9 $\mu\text{g/ml}$
PIC	1.9 $\mu\text{g/ml}$
TAT	0.4 $\mu\text{g/l}$
soluble fibrin monomer	1.7 $\mu\text{g/ml}$
AT-III	97
Protein C	83%
Protein S	123%

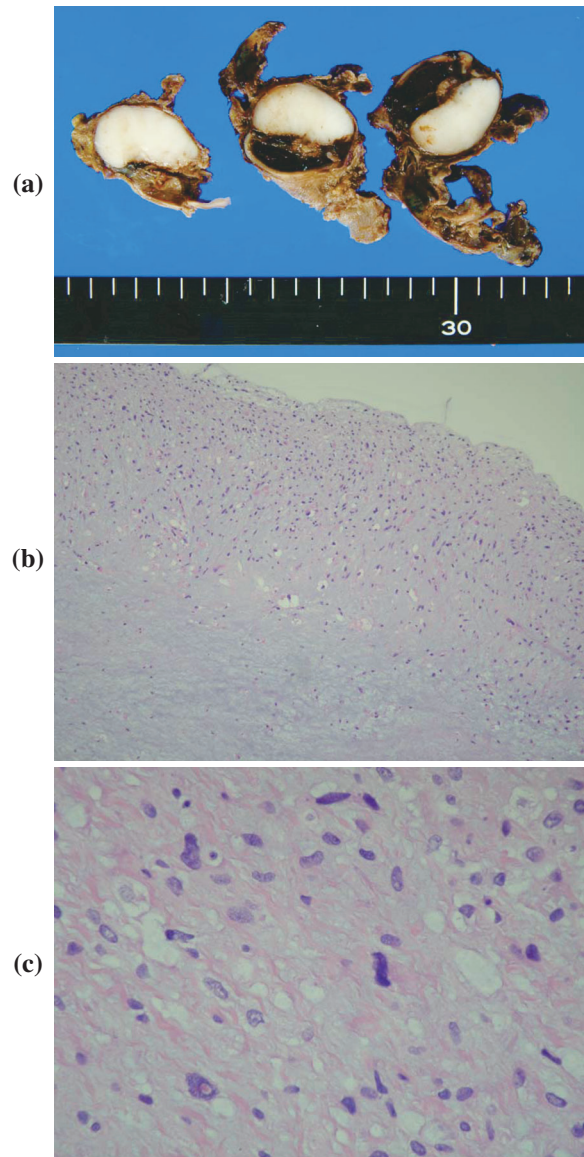
helpful for differentiating a tumor mass from a thrombus (3-5), by showing an enhancement of an intraluminal filling defect with Gd-DTPA (Fig. 3). To confirm whether or not a tumor was located in the pulmonary artery, we performed FDG-PET CT (fluorodeoxyglucose positron emission CT), showing an RI uptake in the mass (Fig. 4), which thus indicated the presence of a neoplasm in the pulmonary artery. FDG-PET CT and enhanced CT showed no primary lesion elsewhere, and we performed bronchoscopy to rule out invasion from a bronchogenic tumor into the pulmonary artery, which detected no tumor. These findings indicated that the tumor arose primarily from the pulmonary artery. Thereafter, a surgical resection of the pulmonary artery and the right lung was performed for both a definite diagnosis and therapy. First, a yellowish-white firm gelatinous polypoid mass, measuring 6 cm long and 4.5 by 1.5 cm in diameter, was removed from the pulmonary artery, and then the pulmonary artery was resected which revealed a residual tumor, measuring 2.5 by 1.5 by 1 cm in size, filling the artery with a dark-red thrombus (Fig. 5a). Histologically, the tumor consisted of a fascicular or ill-arranged proliferation of atypical spindle cells in a myxoid or fibromyxoid background, with hypocellular and hypercellular areas (Fig. 5b, c). The cells were hyperchromatic with a small to moderate amount of eosinophilic clear cytoplasm. The nuclei showed mild to severe atypia or some pleomorphic bizarre nuclei. There were few mitoses. There were no foci of more differentiated sarcomas such as osteosarcoma, chondrosarcoma, rhabdomyosarcoma, leiomyosarcoma, or angiosarcoma. The tumor extended distally into bronchus at the hilum of the right lung. Most of the tumor extended along the intima and focally showed compressing growth into adventitia. There was no invasion of the tumor into the lung parenchyma. Paraffin sections of the tumor were immunohistochemically stained using the following antibodies: S-100 protein, EMA, keratins (34  $\beta\text{E12}$ , and AE 1/3), desmin, actin,  $\alpha$ -smooth muscle



**Figure 3.** Chest MRI showing a slightly high intensity mass in the pulmonary artery on T2-weighted image (a), with a low intensity on the T1-weighted image (b). An enhancement effect is found on the T1-weighted image (c).



**Figure 4.** FDG-PET CT. The RI uptake of a filling defect in the pulmonary artery is shown.



**Figure 5.** Pathology. a. A whitish mass filled in the pulmonary artery with a thrombus. b. The tumor consisted of an ill-arranged proliferation of atypical spindle cells in a myxoid background with hypercellular and hypocellular zones (H&E; original magnification  $\times 100$ ). c. The cells were hyperchromatic with a small to moderate amount of eosinophilic to clear cytoplasm. Some cells showed bizarre pleomorphic nuclei (H&E; original magnification  $\times 400$ ).

actin ( $\alpha$ SMA), CD31, CD34, and D2-40. The cells were non-reactive to all antibodies, except for very focal positivity with desmin and  $\alpha$ SMA. Based on the clinical, morphological, and immunohistochemical findings, the tumor was diagnosed to be PAS, or intimal sarcoma of the pulmonary artery.

She was discharged after the operation, and thereafter was regularly followed up on an outpatient basis. The tumor has not recurred for 8 months after the operation.

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## Discussion

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This case was diagnosed to be PAS based on the findings of a complete medical examination. Primary PAS is rare, and only 150 cases have been reported in the world. Histologically, PAS have been subclassified as follows: undifferentiated, 34%; fibrosarcoma or fibromyxosarcoma, 21%; leiomyosarcoma, 20%; rhabdomyosarcoma, 6%; mesenchymoma, 6%; chondrosarcoma, 4%; angiosarcoma, 4%; osteosarcoma, 3%; malignant fibrous histiocytoma, 2% (6, 7). However, the histologic subclassification of these tumors does not seem to be useful either clinically or prognostically (7); most of the tumors are undifferentiated sarcomas with focal fibroblastic or myofibroblastic differentiation (WHO classification Tumors of the lung, pleura, thymus and heart, IARC, 2003, and/or Tumors of soft tissue and bone 2002). Most reports have shown that PAS arise within the pulmonary trunk or pulmonary valve region, and therefore they are frequently associated with multicentric origin in the outflow tract of the right ventricle (7). Most reports distinguish between sarcomas of the pulmonary parenchyma or of the heart which secondarily invade the pulmonary artery, and therefore arise primarily from the intima of the pulmonary artery. In the present case, the tumor developed from inside the pulmonary artery while compressing the adventitia, however, it demonstrated no contact with either the bronchus or heart, thus indicating pulmonary artery origin. The presenting symptoms with PAS are similar to those of patients with other pulmonary vascular diseases, including progressive dyspnea, chest pain, cough, and hemoptysis (6, 7). Progressive weight loss, anemia, and fever, which are not uncommon in pulmonary vascular diseases, are also observed in patients of PAS. The present case occasionally showed fever and bloody sputum, but not hemoptysis. Wackers et al (8)

suggested hemoptysis to be indicative of tumor emboli; however, we regarded the bloody sputum in this case to be derived from the pulmonary infarction, because no tumor cells were observed in the bronchus or the resected lung parenchyma specimens.

The diagnosis is often not suspected prior to surgery, which is often undertaken based on a different preoperative diagnosis such as bronchogenic carcinoma or chronic pulmonary thromboembolism. Certainly, in the present case, a pulmonary artery thromboembolism was initially suspected, however, no venous thrombus was detected, and the abnormalities of coagulation survey were very mild. Chest CT did not show any characteristic signs of chronic pulmonary thrombosis (9), and anti-coagulation therapy (heparin sodium 15000 units/day) for the prophylaxis of secondary thrombosis due to an impaired pulmonary artery circulation showed no improvement in the filling defect. In addition, enhanced MRI findings and PET-CT were very useful for differentiating the tumor from a thrombus. Although there are few reports which had shown usefulness of FDG-PET imaging for the diagnosis of pulmonary artery tumor, this case shows the usefulness for making a global assessment of an atypical filling defect in the pulmonary artery.

Although a rare entity, we should nevertheless consider the possibility of pulmonary sarcoma when evaluating patients with atypical presentations of pulmonary thromboembolism and pulmonary infarction, especially if they are associated with symptoms such as weight loss, fever, and anemia, if atypical results of coagulation surveys for pulmonary thromboembolism are observed. In addition, the disease should be considered if long-term anticoagulation therapy is not successful. We were able to successfully perform a complete resection based on these above described points together with the use of both MRI and FDG PET-CT, which were found to be useful diagnostic modalities.

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