

## Case report - Pulmonary Primary pulmonary artery sarcoma

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### Abstract

Primary pulmonary artery sarcoma is an uncommon tumor. We report a case of a 73-year-old male patient with a two-week history of palpitations and shortness of breath, aggravated for two days and was believed to be pulmonary hypertension. Emergency heart ultrasound after admission presented a massive pulmonary embolism in the pulmonary artery. The patient's condition was successfully managed with urgent pulmonary artery embolectomy. The patient demonstrated improvement in hemodynamics after the operation. Histologic and immunohistochemical assays were performed and a diagnosis was made as primary pulmonary artery sarcoma arising from the left pulmonary artery. Resection of the tumor is recommended for the treatment of this rare malignant tumor. The corresponding chemotherapy, follow-up and prognosis are described as well in this case report.

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**Keywords:** Pulmonary artery; Sarcoma; Embolism

### 1. Introduction

A 73-year-old male with palpitations and shortness of breath for two weeks and symptoms aggravated in the last two days, was admitted on 27 July 2006. He was in a good health condition before then and had no traceable disease history. Physical examination revealed that the patient could not tolerate slight movement, even with the help of the oxygen mask, and a systole murmur in the pulmonary artery valve area was heard. Two-dimensional echocardiography demonstrated a 4.8 cm×5.2 cm mass in the pulmonary artery which was shown to be almost completely obstructed (Fig. 1). Therefore, it was diagnosed as pulmonary embolism initially.

Based on these examinations, we performed an emergency operation for this gentleman. After the sternotomy and pulmonary artery incision, the patient was found to have a solid tumor in the main pulmonary artery, stemming from the base of the main pulmonary artery (Fig. 2, and histology is shown on the bottom right corner). Under the cardiopulmonary bypass for 39 min, the tumorous mass was resected completely. The patient recovered well. There was no detectable tumor mass in the pulmonary artery by echocardiography, and no bone metastasis was found by ECT scanning either. The patient was therefore discharged on day eight after the operation.

The pathologic features of the resected mass were characterized. Macroscopically, the mass was approximately 5 cm×4 cm×8 cm in size with almost intact capsule. In section, myxoid and solid white-gray areas were observed.

H&E-staining of the slides demonstrated cystic and tubular structures lined by single to double cuboidal cell layers under microscopy. Limited to our condition, only some of the immunohistochemical analysis was performed and the results were as follows: S100 (+), NF (neurofilament) (+), Des(+), SMA(-), CD117(-). The final diagnosis given by our pathology department was primary pulmonary artery sarcoma, malignant fibrous histiocytoma (MFH) was much likely.

The patient received the following chemotherapies: cyclophosphamide (IFO) dose: 3.0 g (day 1-3) and 5-(3, 3-dimethyl-1-triazeno) imidazole-4-carboxamide (DTIC) 390 mg (day 1-4). During the first cycle of chemotherapy, the patient could tolerate treatment in general, however, the patient and the family refused further chemotherapy after that. Follow-up chest CT-scanning and heart ultrasonic demonstrated no re-occurrence of tumor in the lung and the main pulmonary artery. After six months, however, tumorous masses were found in multiple organs, including right adrenal gland, bilateral chest walls, and liver, which is indicative of occurrence of metastasis. The patient died of the brain metastasis in March 2007.

### 2. Comment

Primary pulmonary artery sarcoma is an uncommon malignant tumor of the cardiovascular system. Mandelstamm first described this disease in 1923. Since then, around 125 cases were reported with a slight predilection for women [1] and appear in an age range of 13-86 years, with the majority of cases occurring when the patient is 45-55 years [2].

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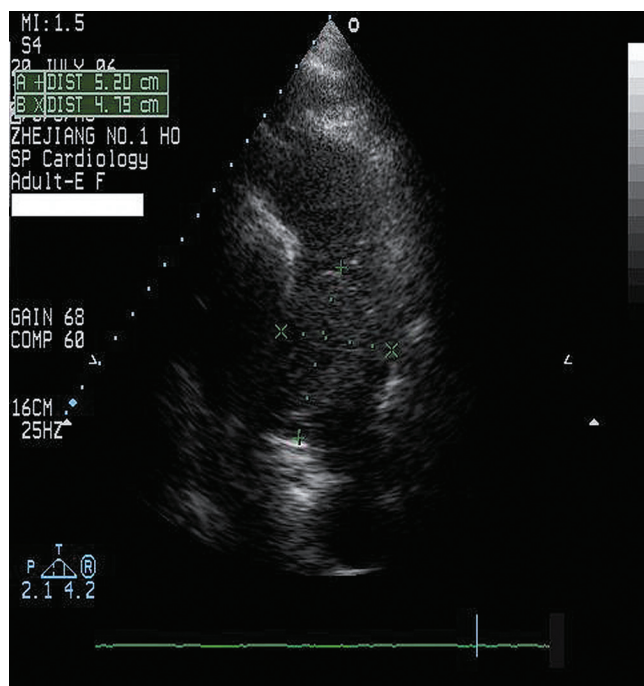


Fig. 1. The mass is shown in the main pulmonary artery before operation by ultrasonic image.

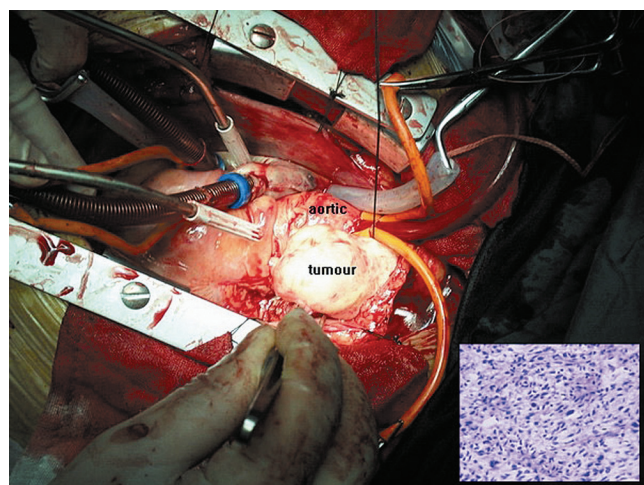


Fig. 2. A solid tumor was found in the main pulmonary artery during the operation and the histology of the tumor (H&E-staining,  $\times 400$ ) is shown in the bottom right corner of this picture.

Symptoms of this tumor can vary depending on the tumor histology, size and location. The most common symptoms [3] include dyspnea, cough, chest pain, malaise, and anorexia. Occluded circulation induced coagulation abnormal, arrhythmias, pericardial effusion tamponade and signs indicative of neoplasia, such as weight loss, clubbing, anemia, and an elevation in the erythrocyte sedimentation rate may occasionally be found in the admission. Systolic murmur of the pulmonary artery valve area origin and signs of dysfunctional right ventricle can be frequently observed in physical examination.

Usually, a chest radiograph can detect hilar enlargement and the echocardiography can locate the mass in the vessels.

In clinical practice, a primary pulmonary artery tumor is extremely rare, whereas the prevalence of pulmonary embolism is fairly high. Due to the fact that their manifestations and the heart ultrasonic findings are similar, the primary pulmonary artery tumor is often miscellaneous with the pulmonary embolism. The diagnosis of pulmonary artery sarcoma is often made right during the operation when the mass is thought to be chronic pulmonary emboli originally.

As for clinical tools to diagnose this tumor, despite that chest CT-scan and cardiac MRI are helpful anatomically, PET scan recently has been proven to be more potent to differentiate these two diseases and to characterize the features of tumor mass in the pulmonary artery [4].

Chemotherapy and radiotherapy are unable to rapidly relieve pulmonary artery obstruction and prevent heart failure. Due to the limitations of chemotherapy or radiotherapy, surgical resection has been developed to be the most beneficial alternative for the treatment of pulmonary tumor. In some cases, resection requires replacing some of the pulmonary root with pulmonary allograft [5], and pneumonectomy is sometimes needed as well.

The prognosis is poor, with short survival time reported: less than two months for patients receiving no surgical treatment and approximately 10 months for those treated surgically – albeit in some cases survival of longer than 10 years has been reported [6].

Pathologically, pulmonary artery sarcoma can be classified into two groups: intraluminal and intramural, according to its occurrence sites [7]. The more common intraluminal form, also known as intimal sarcoma, usually originates from the intimal layer and proliferates as polypoidal masses in the lumen of the vessel.

Radiation therapy and chemotherapy has been recommended for some patients with pulmonary artery sarcoma, in which chemotherapy is preferred, although the effective long-term control strategies remain to be identified. The most widely accepted regimen, used for many sarcomas, includes doxorubicin and ifosfamide [1]. The best efficacy is obtained when dose-intensive chemotherapy is applied. In addition, extra supportive care to prevent the potential complications of chemotherapy is critical. Based on the previous reports, prognosis of this tumor varies with a survival time ranging from several months to decades of years.

In conclusion, when a pulmonary embolism has been diagnosed, and the anticoagulant treatment begins, extra attention for pulmonary artery sarcoma should be kept in mind for those patients who have unilateral pulmonary artery occlusive disease with no evidence of positive hypercoagulability tests and no history of thromboembolism. In those cases, an angiographic-guided biopsy from the intra-arterial occlusive material should be performed, and PET scan may be helpful as well. As for treatment, surgical resection is considered to be most effective and preferred, combining adjuvant chemotherapy and radiotherapy if the patient can tolerate. In summary, more case reports and analysis are required for further understanding the poten-

tial pathogenesis as well as development of a comprehensive control strategy of this disease.

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