Pulmonary artery sarcoma mimicking pneumonia: case report and literature review

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Abstract: Pulmonary artery sarcoma is a highly malignant and very rare tumor with a high rate of misdiagnosis. We present a case of a patient who complained of recurring fever for six months. He was diagnosed with pneumonia in a tertiary hospital in Nanjing and treated for more than one month, and was admitted with an initial diagnosis of pneumonia to our hospital. Pulmonary CT Angiography (CTPA) demonstrated pulmonary embolism. Until positron emission tomography-computed tomography (PET-CT) was performed, the patient was suspected to have pulmonary artery sarcoma (PAS). According to the histopathology from total left lung resection, he was eventually diagnosed with PAS and recovered very well. The prognosis of PAS depends very closely on early surgical treatment, and therefore physicians must be aware of PAS and grasp the best treatment opportunity.

Keywords: Pulmonary artery sarcoma, pulmonary thromboembolism, pneumonia

Introduction

Pulmonary artery sarcoma (PAS) is a highly malignant tumor that originates from the pulmonary arterial intima. PAS is very rare. To date, there are only 400 reports of the disease in the literature, mostly as case reports [1]. The onset of PAS is occult, the clinical symptoms are lacking in specificity, and the rate of misdiagnosis is extremely high. The clinical manifestations of PAS are often very similar to those of pulmonary thromboembolism (PTE), such as progressive dyspnea, chest pain, cough, fatigue, hemoptysis and syncope, while symptoms such as fever, dizziness, and palpitations are rare [2].

Case presentation

A 52-year-old man was admitted to Affiliated Hospital of Hangzhou Normal University (Hangzhou, China) on March 22, 2017, with the complaint of recurring fever for six months. He was diagnosed with pneumonia in a tertiary hospital in Nanjing and treated for more than one month. The patient had no history of hypertension, heart disease, or tumor. There were no significant findings on physical examination. Pulmonary CT (January 1, 2017; January 14, 2017) demonstrated left lung infection, inflammation of both lungs, and multiple mediastinal lymph nodes (Figure 1).

Hospital laboratory test results revealed white blood cell count (WBC) 6.63 × 10^9/L, neutrophil percentage (NEU%) 63.3%, high-sensitivity C-reactive protein (CRP) 153.6 mg/L and procalcitonin < 0.25 ng/ml, Mycoplasma pneumoniae antibody positive (1:1280); D-dimer 0.27 mg/L. Tumor indicators: carcinoembryonic antigen (CEA) 1.45 ng/ml, neuron specific enolase (NSE) 21.44 ng/ml; blood lactate dehydrogenase (LDH) 121 U/L; Aspergillus fumigatus IgG antibody quantitative detection was 55 AU/ml (negative), autoantibodies and T cell spot test (T-SPOT test) for tuberculosis infection were also negative. Bone marrow puncture showed hyperplasia of bone marrow hematopoietic tissue.

Pulmonary function tests showed normal lung ventilation function; Cardiac ultrasound showed left atrial enlargement and no abnormalities were found in the pulmonary artery.
was no thrombosis in both upper and lower extremity veins. Gastroscopy showed duodenal ulcer, and superficial gastritis, and enteroscopy showed chronic colitis.

The patient was treated with anti-infective therapy for one week, and his fever continued with a highest body temperature of 38.9°C. On 3-31-2017, pulmonary CT Angiography (CTPA) demonstrated: the left main pulmonary artery and most of the branches were embolized, and the left lung had multiple high-density shadows, suggesting pulmonary embolism.

The diagnosis of this patient was chronic pulmonary embolism. The patient had recurrent episodes of fever for half a year; pulmonary artery sarcoma was considered, and positron emission tomography-computed tomography (PET-CT) was performed.

PET-CT results suggested (Figure 3): 1. The left pulmonary artery lesion size was about 41 mm × 28 mm, the SUV maximum was about 35.5, FDG metabolism was abnormally active, considering malignant lesions, vascular source may be vascular endothelioma; 2. Both lungs had pulmonary-based multiple nodules, some nodules' FDG metabolism was increased slightly, and the lesion was located under the pleura. Chronic inflammation may be reconsidered after anti-inflammatory treatment review unless there was metastasis. The patient was suspected to have pulmonary artery sarcoma.

The patient was transferred to thoracic surgery for total left lung resection. Visible to the naked eye: the whole lung lobe resection specimen was 22*14*6 cm, bronchial diameter 1.8 cm. Blood vessel diameter was 2 cm, and was filled with up to 18 cm of grayish white and grayish red soft tissue. Immunohistochemistry: SMA (-), CD34 (-), CD31 (-), F8 (-), CK-pan (-). Histopathology revealed PAS (Figure 4). The patient recovered well after surgery, with no fever, no chest pain, no difficulty in breathing. The patient did not undergo radiotherapy and chemotherapy, and his condition was stable.
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Discussion

Pulmonary arterial sarcoma (PAS) clinical symptoms and imaging findings lack specificity. PAS laboratory tests also lack specificity. ESR, LDH and hs-CRP may have a certain value for early diagnosis [3]. D-dimer is mostly in the normal range and can be used as a basis for differential diagnosis of PTE [2]. Almost all patients were misdiagnosed with chronic thromboembolism for months or years before pathological diagnosis [2, 4]. In the present case, the patient was misdiagnosed with pneumonia based on PET-CT for several months, and with pulmonary thromboembolism based on pulmonary CTa. Imaging findings plus laboratory tests have a great value in the diagnosis of PAS. Echocardiography can examine the right ventricular outflow tract and main pulmonary artery occupying position, right ventricular diameter, and pulmonary systolic pressure [5]. The manifestation is that inhomogeneous parenchymal filling in the pulmonary arteries is distinct from the uniform echo of fresh thrombus in the pulmonary embolism. It can show the signal filling in the pulmonary artery. It can cause pulmonary hypertension, enlargement of the right heart, and enlargement of the right ventricle.

Figure 3. PET-CT showed: 1. The size of left pulmonary artery stem lesions is about 41 mm × 28 mm, the maximum SUV is about 35.5, FDG metabolism is abnormally active, consider malignant lesions, such as vascular endothelial sarcoma.

after regular review and follow up for more than one year.
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Figure 4. Pathology showed: short-spot or star-shaped stellate cells distributed in pieces, mixed with giant tumor cells; nuclear mitosis is frequent. Tissue degeneration and lymphocyte infiltration, with histiocytes and multinucleated giant cells occurs, and the focal area invades the blood vessel wall, involving the lung tissue. Bronchial margin is negative.

Pulmonary arterial sarcoma progresses rapidly with poor prognosis [13]. Surgical treatment is the first choice for patients without extra-arterial metastasis [14], including pulmonary arterial endarterectomy, tumor and pulmonary artery resection, and pulmonary artery reconstruction, lobectomy, and pneumonectomy. The 5-year survival rate of PAS is extremely low. The average survival time of patients without surgery was only 1.5 to 12 months [15], while surgical treatment could prolong the survival time by 8 to 36 months [1]. Early diagnosis is critical for patients with PAS. To date, the patient benefited from early diagnosis and his condition was stable for more than one year. This case reminds us that although pneumonia is common, we also need to be vigilant against PAS and avoid misdiagnosis and mistreatment.

Conclusions

Pulmonary artery sarcoma is a highly malignant and very rare tumor with a high rate of misdiagnosis. The clinical manifestations of PAS lack specificity, so it may be misdiagnosed as pulmonary thromboembolism or as pneumonia. Clinicians need to strengthen their understanding of the disease, reduce misdiagnosis, and improve patient outcomes.

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Disclosure of conflict of interest

None.

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