Case Report

Primary pulmonary artery sarcoma with intrapulmonary metastases based on PET/CT imaging: a case report and literature review

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Abstract: Primary pulmonary artery sarcoma is an extremely rare and highly aggressive malignant tumor of cardiovascular system. It is usually misdiagnosed as pulmonary thromboembolism due to its atypical clinical features and similar clinical symptoms. Different from published reports, our case received both enhanced CT and ^{18}F-FDG PET/CT examination before the pathologic result, and lung metastases had already occurred at the time of diagnosis. We herein reported a case of 41-year-old female patient who suffered from cough and chest tightness for more than a month. Laboratory examination indicated that both blood routine and tumor markers were within the normal range, and only the D-dimer slightly elevated. Contrast-enhanced chest computed tomography showed right pulmonary artery lesion and multiple nodular located right upper lung, the lesion was mild heterogeneous enhancement. No obvious abnormalities were found in deep vein of bilateral lower extremities on ultrasonography. In order to confirm the nature of these lesions, PET/CT scan was performed, which revealed stripe hypermetabolism in right pulmonary artery and nodular hypermetabolism in right upper lung, and the rest of the whole-body PET imaging were negative, a diagnosis of primary pulmonary artery malignancy with pulmonary metastases was made, and pulmonary thromboembolism was ruled out. Biopsy of right pulmonary lesions was performed and histopathological examination indicated pulmonary artery sarcoma. She only received palliative conservative medical treatment because the disease was late stage according to the tumor-node-metastasis (TNM) staging system, and did not acceptable surgical treatment, and was in good health during recent follow-up. Our study suggested that 18F-FDG PET/CT image is a good approach for the diagnosis of pulmonary artery sarcoma and could provide adjunct value for further treatment.

Keywords: Pulmonary artery sarcoma; 18F-FDG; PET/CT; intrapulmonary metastases; case report

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Introduction

Sarcoma is a malignant tumor derived from mesenchymal tissue. Pulmonary artery sarcoma (PAS) is a rare site of sarcoma and fewer than 400 cases have been reported in the literatures, which only accounts for approximately 0.001–0.003% of all sarcomas (1,2). Consequently, PET/CT imaging used for PAS is even rarer, only a few case reports have described these lesions, but all these cases were misdiagnosed as pulmonary embolism or pneumonia at initial diagnosis with the imaging manifestations of FDG high uptake on PET/CT, and finally confirmed as PSA by postoperative histopathological results (3-5). Herein we

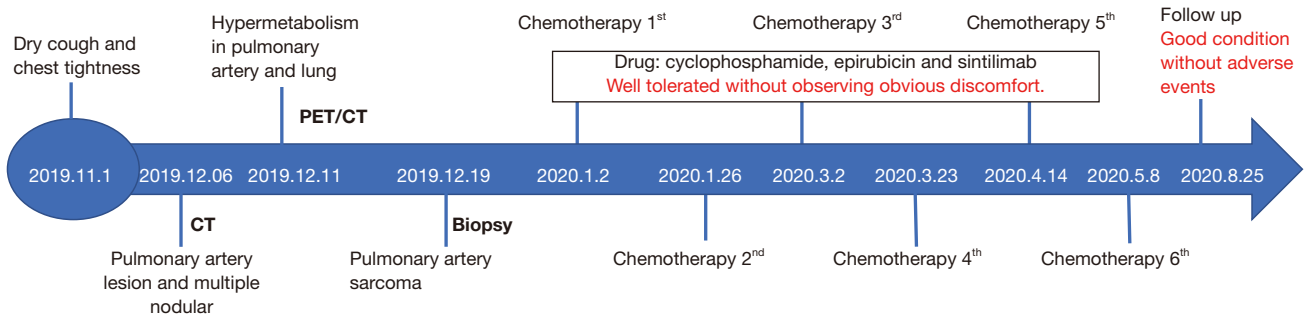

Figure 1 Time of clinical history for pulmonary artery sarcoma. PET indicates positron emission tomography; CT indicates compute tomography.

present a case of 41-year-old man with initial diagnosis of PAS. She underwent a biopsy of the right lung and histopathology examination revealed pulmonary artery spindle cell sarcoma, she did not undergo operation because the disease is late stage, and received chemotherapy and immunotherapy. This rare case showed hypermetabolic within the right pulmonary artery accompanying multiple metastases in the right upper lung on PET/CT, indicating malignant lesions. The time-line of the clinical history for this patient was shown in *Figure 1*.

We present the following article in accordance with the CARE reporting checklist (available at http://dx.doi.org/10.21037/apm-20-630).

Case presentation

The study was approved by committee ethics board of the first affiliated hospital of university of science and technology of china (No. 2018-ky028). All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

A 41-year-old women presented with dry cough and chest tightness for more than a month. Plain CT scan of the chest at a local hospital revealed a soft mass in the right hilar and multiple nodules in right upper lung, and she was referred to our hospital for further treatment. Contrast enhanced CT indicated pulmonary artery filling defect with multiple nodular lesions in the right lung, color doppler echocardiography showed moderate pulmonary hypertension and decreased left ventricular diastolic function, and no abnormalities were seen in deep venous of both lower extremities with sonography, a diagnosis of pulmonary artery sarcoma with right lung metastatic tumors was made. No enlargement of liver, spleen, and superficial lymph nodes were detected. Her initial blood pressure was 108/70 mmHg, and heart rate was 89 beats per minute, she took approximately 20 breaths per minute, and the CO_2 binding capacity was 23.20 mmol/L (normal range: 20.00–30.00). laboratory tests showed neuron-specific enolase slightly elevated to 23.09 ng/mL (reference range: 0.00–17.00 ng/mL), the D-dimer slightly elevated to 1.28 μg/mL (reference rang: 0.01–0.55 μg/mL), tumor marker AFP, CEA and CA19-9 were all within normal limits, blood routine results were also in normal range. She had a history of cesarean section operation two month ago, and denied any history of infectious diseases such as hepatitis, tuberculosis, and malaria, and had no history of hypertension, heart disease, cerebrovascular disease, or psychosis, and no history of family tumor or genetic disease. To further assess the lesions of within right pulmonary artery and right lung, PET/CT scan was then performed. The lesion within right main pulmonary artery and nodules in right upper lung manifested increased FDG uptake [maximum standardized uptake value (SUVmax) 12.7, *Figure 2*], a pulmonary artery sarcoma with intrapulmonary metastasis were diagnosed, meanwhile, she was also diagnosed with pulmonary hypertension. Then right lung nodule biopsy was performed under CT-guided, histopathology examination showed malignant tumor mainly composed of spindle cells, and immunohistochemical staining was positive for Vim, TTF-1, CD34, and Bcl-2, and negative for CK7, Syn, CgA, and CD10, and the Ki-67 level was approximately 80%, which suggested spindle cell sarcoma (*Figure 3*).

The clinical stage of the tumor was T4N0M0 and the KPS score was 80. After a multidisciplinary consultation, she received conservative comprehensive treatment

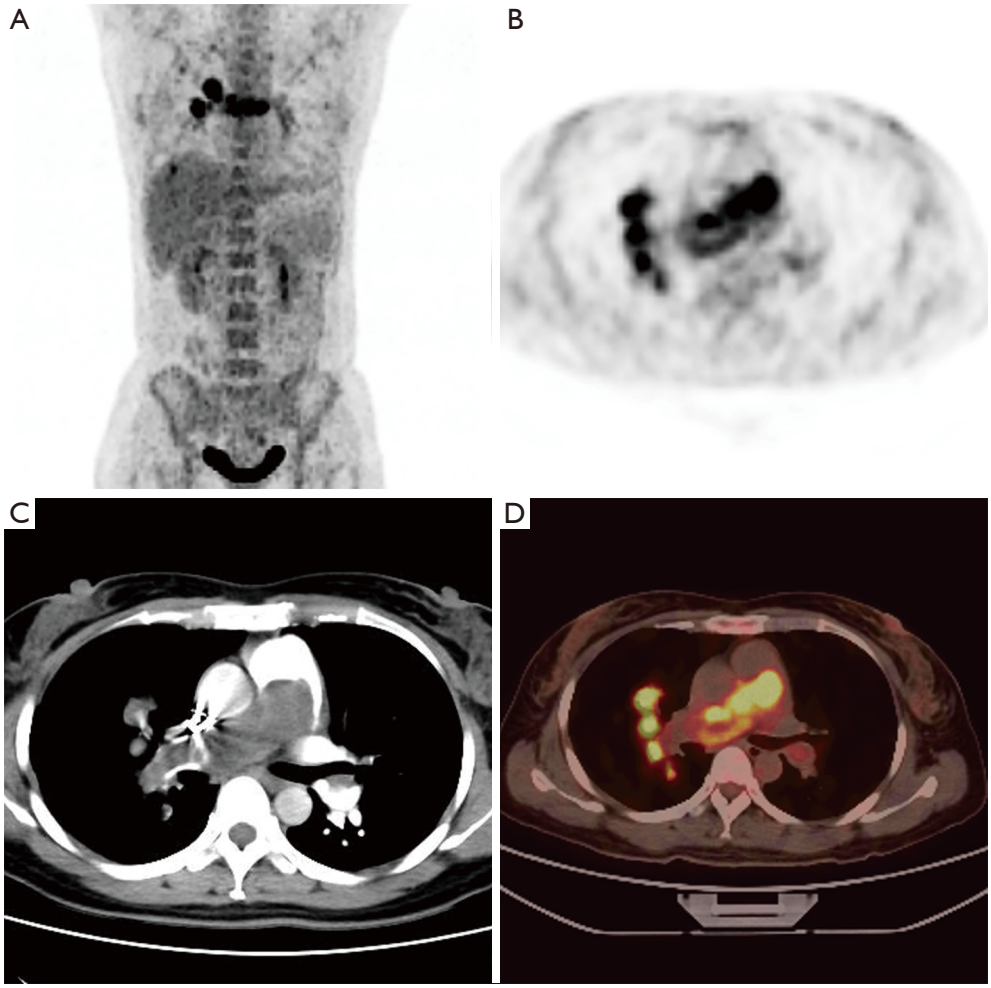

Figure 2 Radiographic examinations using CT and PET/CT. (A) Nodular hypermetabolism lesions were seen in mediastinum and superior lobe of right lung on PET image; (B) a striped hypermetabolic focus in the main right pulmonary artery and multiple hypermetabolic foci in superior lobe of right lung were observed on transverse PET image; (C) filling defects were seen in right main and the branches of pulmonary artery on axial contrast enhanced CT; (D) multiple hypermetabolic foci in right upper lung and striped FDG avid in the main right pulmonary artery were seen on axial fusion image. PET, indicates positron emission tomography; CT, indicates compute tomography; FDG, fluoro-deoxy-glucose.

instead of surgery or radiotherapy. And she was willing to receive chemotherapy plus immunotherapy. she received chemotherapy with cyclophosphamide 1.2 g per day and epirubicin 80 mg per day for 5 courses of chemotherapy. At the same time, she received immunotherapy with sintilimab 100 mg per day 5 courses of chemotherapy, and is in the hospital for six-cycle chemotherapy, no obvious adverse reactions appeared during chemotherapy. She was in good health during 6-month of voluntary follow-up with color doppler echocardiography, and nothing adverse or unexpected happened. She agreed to have her clinical data and images published in journals for use in medical academic exchanges.

She underwent a total of 6 cycles of chemotherapy so far, and his symptoms were significantly improved compared with those before treatment. Symptoms of chest distress were mild, shortness of breath was significantly improved, and he had no other complaints of discomfort.

Discussion

Primary pulmonary artery sarcoma is a rare malignant

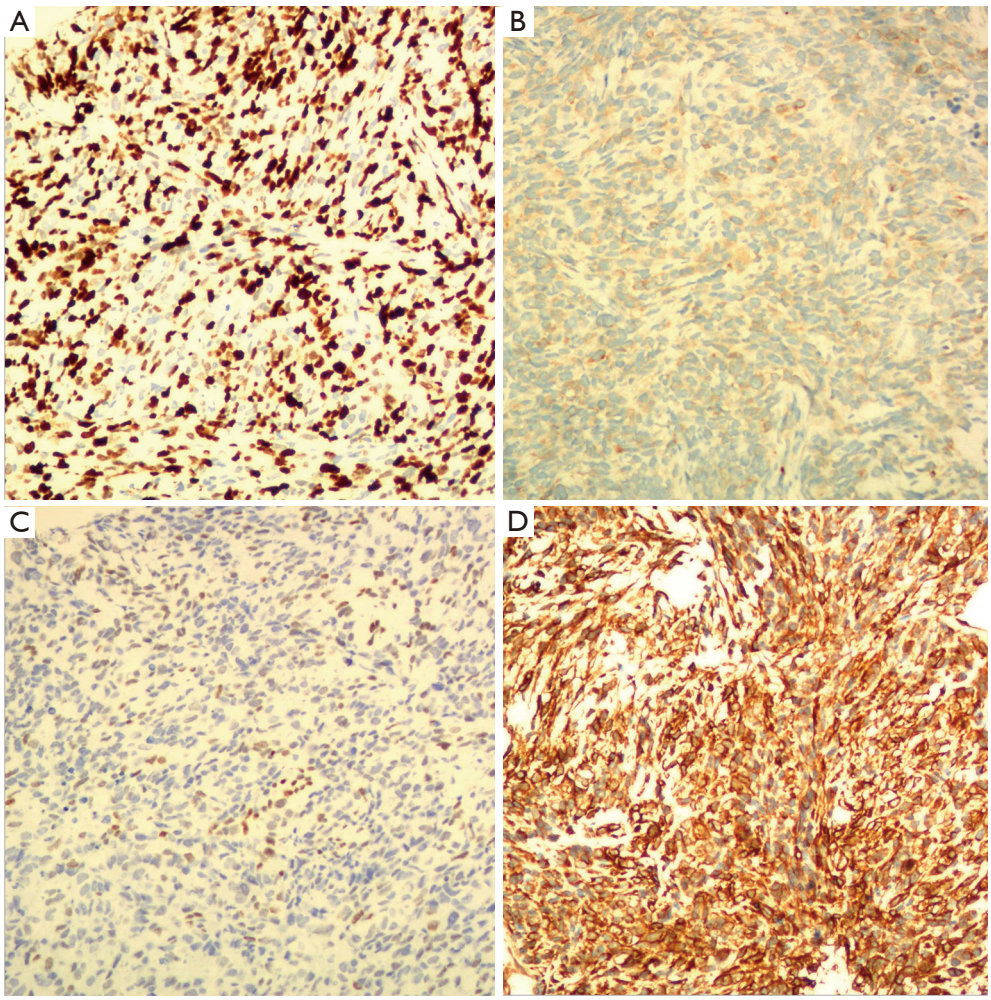

Figure 3 Histopathological analysis. (A) IHC staining was positive for Vim; (B) IHC was positive for TTF-1 (weak); (C) IHC was positive for Bcl-2; (D) the Ki-67 proliferation index was approximately 80%. All magnifications were ×200. IHC, immunohistochemistry.

tumor originating from the intima or subintima of the pulmonary artery, less than 400 cases have been reported since it was first described in 1923 by Mandelstamm (6,7). The exact etiology is unclear, and the incidence is about 0.001% to 0.003% (2), the disease is more occult at early stage, the non-specific symptoms include chest pain, cough, dyspnea, hemoptysis or sputum with blood, syncope and asymptomatic. Because clinical manifestations of pulmonary artery sarcoma lack specific symptoms and it is difficult to make accurate diagnosis at early stage, most of the patients were misdiagnosed as pulmonary thromboembolic disease before surgery (8-10). After repeated thrombolytic therapy failed, and the diagnosis of pulmonary artery sarcoma was made during surgical treatment. The imaging findings of echocardiography, and pulmonary vascular magnetic resonance imaging were similar to those of pulmonary embolism, the 'wall eclipsing sign' on pulmonary artery computed tomography angiography is typical characteristic for diagnosis of PAS (1). Pulmonary artery sarcoma could manifest increased FDG uptake on PET/CT imaging, however, pulmonary artery thromboembolism usually shows radioactive defect, which is important for the differential diagnosis between pulmonary artery sarcoma and pulmonary thromboembolism, and ^{18}F-FDG PET/CT have been shown useful in the diagnosis of pulmonary artery sarcoma and also could contribute to an accurate diagnosis for determining pulmonary artery malignancy (4,11,12). In addition, ^{18}F-FDG PET/CT could provide whole body condition through once examination, and whether the tumor has distant metastasis. Accordingly,

it could provide useful information for tumor diagnosis and staging. However, not all pulmonary artery sarcomas show hypermetabolic on ^{18}F-FDG PET/CT imaging, and the minority of pulmonary artery intimal sarcoma can present hypometabolism at ^{18}F-FDG PET/CT image, which is easily misdiagnosed as pulmonary embolism (8). The main imaging features of pulmonary artery sarcoma include filling defects with hypermetabolism on PET/CT images, expansion the artery, metastasis to the lungs or mediastinum, and delayed enhancement on CT image, which are different from pulmonary artery blood thrombosis. Although there was no standard TNM staging system at present, optimal treatment regimens are adopted for different staging tumor. Pulmonary artery sarcoma is a rare and highly lethal disease, and its diagnosis and optimal treatment strategy are not conclusive. Surgery is preferred treatment choice for pulmonary sarcoma up to now. Patients will benefit from early diagnosis and complete surgical resection, and postoperative adjuvant treatment of management may prolong the patient's survival (13). Patients with untreated pulmonary artery sarcoma would be a median survival of 6 weeks (14). Although the surgery and the newer upcoming treatment strategies may prolong the median survival of patients, the long-term prognosis remains extremely poor (15).

In our case study, a complete auxiliary examination was performed with pulmonary artery angiography and ^{18}F-FDG PET/CT imaging for our case, pulmonary angiography suggested mild enhancement of pulmonary artery lumen lesions, and pulmonary embolism was first considered. ^{18}F-FDG PET findings indicated that pulmonary artery lesion accompanied by multiple nodules in the right upper lung, a pulmonary artery malignancy accompanied by right metastasis was made, and pathologic findings of right lung nodule revealed sarcoma. The disease was at an advanced stage, so surgical treatment could not be performed, Chemotherapy plus immunotherapy is first-line treatment for pulmonary artery sarcoma, to our knowledge, sintilimab used as a treatment for PAS has not been reported previously. Up to now, the patient is in good condition and is under follow-up. The limitations of our study are that we cannot predict eventual survival while patients are still being treated and followed up. Chemotherapy combined with immunotherapy may be a good treatment approach for advanced pulmonary artery sarcoma, and patient would benefit from it. When a lesion in the pulmonary artery cavity demonstrates hypometabolism on PET imaging, it is easily to rule out pulmonary embolism due to its hypometabolism on PET image. The case presented with pulmonary arterial luminal mass, enhanced CT indicated mild enhancement of the lesion, and the size of the lesion did not change after anticoagulant treatment. The lesions in the arterial luminal indicated hypermetabolism accompanied by hypermetabolism nodules in the right lung on ^{18}F-FDG PET/CT imaging. Pulmonary artery malignancy with intrapulmonary metastasis was first considered, and pulmonary thromboembolism could be excluded. Therefore, when there were nodular or striped hypermetabolic foci in the pulmonary artery cavity at ^{18}F-FDG PET imaging, primary pulmonary artery sarcoma should be taken into consideration firstly.

Conclusions

Primary pulmonary artery sarcoma should be firstly taken into consideration when a nodular or striped hypermetabolism of intraluminal lesions of pulmonary artery on PET/CT image. Other findings include metastasis to lung or mediastinum may provide adjunct diagnostic value for pulmonary artery sarcoma.

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Footnote

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/apm-20-630). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are

appropriately investigated and resolved. The study was approved by committee ethics board of the first affiliated hospital of university of science and technology of china (NO.2018-ky028). All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

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