Radiologic findings of primary pulmonary angiosarcoma
A case report
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Abstract
Rationale: Primary pulmonary angiosarcoma is a rare disease. Here, we report the case of primary pulmonary angiosarcoma diagnosed computed tomographic pulmonary angiography (CTPA) and discuss its specific imaging characteristics.

Patient concerns: A 46-year-old man was admitted for cough and shortness of breath. Thoracic CTPA images demonstrated a high-attenuation lesion surrounding by a halo sign in upper lobe of right lung, and the dilated vessel was also seen in lower lobe of right lung. The sign of “hillside sign” was observed on CTPA.

Diagnoses: It was diagnosed with primary pulmonary angiosarcoma.

Interventions: Right thoracotomy and right upper lobe lobectomy were performed.

Outcomes: Five years later, the patient dead of complete occlusion of the pulmonary artery owing to tumor recurrence.

Lessons: Although primary pulmonary angiosarcoma is a rare disease with atypical early clinical symptoms, and it is often misdiagnosed as pulmonary embolism and pulmonary infection. Therefore, it is important to recognize the CTPA imaging characteristics of primary pulmonary angiosarcoma and Surgical resection should be performed to prolong the patients’ lifetime.

Abbreviations: CRP = C-reactive protein, CT = computed tomography, CTPA = computed tomographic pulmonary angiography, GGO = ground-glass opacity, WBC = white blood cell.

Keywords: angiosarcoma, computed tomographic pulmonary angiography, computed tomography, pulmonary angiosarcoma

1. Introduction
Pulmonary angiosarcoma is a rare malignant neoplasm of lung including primary and metastatic tumor. The incidence of primary pulmonary angiosarcoma is about 0.001% to 0.030%. It mainly occurs in middle-aged male. The postoperative survival time is only about 12 months. The previous literatures are mostly about its pathology. Little is known about the imaging features of primary pulmonary angiosarcoma on computed tomography (CT) in the English literature to date. The purpose of this case report is to describe the specific imaging features of primary pulmonary angiosarcoma found in the patient.

2. Case report
A 46-year-old man was hospitalized with a 1-month history of cough. He had no personal or family history of respiratory diseases. Physical examination demonstrated no significant abnormalities. CT images showed patchy exudative lesion in the upper lobe of right lung and the thickening right inferior pulmonary artery (Fig. 1 A,C). Laboratory examination revealed an elevated C-reactive protein (CRP, 64mg/L) and D-dimer (500ng/mL); the white blood cell (WBC) count was 11.47 × 10^9/L, and tumor marker level was normal. The case was diagnosed as infection and pulmonary embolism. Then, he was given anti-inflammatory and anticoagulant treatment, and he was discharged after the cough symptom completely disappeared. Two months later, the patient’s symptom appeared again. Then, he was followed up with CTPA. The primary lesion of right lung had been basically absorbed and a new lesion was found in right upper lobe (Fig. 1B). The thickening right inferior pulmonary artery was significantly enlarged and distorted (Fig. 1D). Mediastina window images demonstrated that multiple filling defects were found in the pulmonary conus, main pulmonary artery, and pulmonary branches of the upper right lung, combined with pulmonary artery stenosis (Fig. 1E), and the hillside sign (main lesions were interlinked with each other growing to the pulmonary cavity, with smooth surface and different heights) was also found (Fig. 1F). Then, it was diagnosed as malignant tumor of pulmonary artery. Other related examinations (such as echocardiography, bronchoscopy, sputum pictures, and nuclear medicine) found no abnormalities, and the biopsy was not performed. Right thoracotomy and right upper lobe lobectomy were performed in our hospital. But the tumor in pulmonary...
artery was not completely resected because of its extensive involvement. The intraoperative findings were similar to imaging's performances. Microscopically, part of tumor cells grew with vascular wall (HE, x40), most of the cells were fusiformis with irregular nucleus and significant atypia, and amount of mitotic counts was found in the most cells (HE, x200). Immunohistochemical observation showed that Vimentin and SMA were positive, CD31, CD34, F8, CK, S-100, Desmin, and HMB45 were all negative; the Ki67 index was about 30% (Fig. 2A, B). The patient was eventually diagnosed as primary pulmonary angiosarcoma. The patient had anticoagulant therapy with aspirin in the follow-up, without

Figure 1. Radiological manifestation of primary pulmonary angiosarcoma. (A) Selected axial image of computed tomography thorax showed the halo sign around the high-density lesions in the upper lobe of right lung. (B) Chest computed tomography of pulmonary showed that the primary lesion had been basically absorbed and a new lesion was found in right upper lobe after two months. (C, D) At the same time, the lesion in the lower lobe of right lung with clear boundary demonstrated increasing significantly after 2 months. (E, F) CTPA showed multiple filling defects in the main branches of pulmonary artery and arterial cone with a wide basal, jumping distribution and hillside sign were also found.
chemotherapy and radiotherapy. Five years later, he was dead because of complete occlusion of the pulmonary artery owing to tumor recurrence.

The present study was approved by the Ethics Committee of the first hospital of Jiaxing, and the patient information was anonymized and de-identified before analysis.

3. Discussion

Primary pulmonary angiosarcoma is a difficult disease to diagnose clinically with nonspecific respiratory symptoms. Diagnosis is often late due to low clinical suspicion for the disease, which often leads to a poorer prognosis.[3–5] The most common clinical symptom of primary pulmonary angiosarcoma is hemoptysis, which has been described in previous studies.[6,7] But the initial symptom of our report is cough. Most patients with primary pulmonary angiosarcoma have poor prognosis after surgery excision. The report of longest survival period was 3 years.[8]

3.1. CT findings

The CT findings of primary pulmonary angiosarcoma are various and atypical; Shimabukuro et al.[6] reviewed 31 cases with primary pulmonary angiosarcoma, in which the major CT features included pulmonary nodules (87%), infiltrations (22%), ground-glass opacity (GGO) (13%), pleural effusion (16%), pulmonary nodules surrounded by GGO (9%), and the invasion of other organs (19%). The above finding was mainly about the performances of the lung. Infiltrations were also observed in our case, but we found some differences. Partial pulmonary artery in the right lower lung showed obvious thickening and twisting without intrapulmonary manifestations. We wondered whether it was related to the difference of blood supply between the upper and lower lobes of lung. However, more reports are needed to confirm this speculation. Multiple filling defects were found in the main pulmonary artery and arterial cone on CTPA images. The major CT features of multiple filling defects included smooth surface, narrow lume, wide base protruding into the cavity, and a hillside sign, which has not been reported in the past literatures. In addition, the lesions displayed significant enhancement. The lymphadenopathy of right hilar in our case was also observed. In the end, the lymphonodus were inflammatory confirmed by pathology. The existence of pleural effusion in primary pulmonary angiosarcoma is rare. Pleural effusion was also not observed in our case.

3.2. Differential diagnosis

The etiology of primary pulmonary angiosarcoma is not clear, and there is no difference in male and female patients. Predisposing factors include thorium dioxide exposure, polyvinyl chloride, postmastectomy, postirradiation states, and chronic empyema for pleural angiosarcomas.[9] Primary pulmonary angiosarcoma is difficult to distinguish from other lung diseases owing to nonspecific respiratory symptoms of early stage. It is mainly distinguished from the following diseases on the image.

The pulmonary embolism demonstrates multiple filling defects found in the pulmonary artery; it mostly occurs in the lower lobe lung with no thickening of the pulmonary artery and enhancing filling defect.

Primary pulmonary angiosarcoma is also required to be differentiated from pulmonary vascular malformations; the dilatation and tortuosity of pulmonary artery are found in the latter without filling defect in the cavity.

In addition, it has to be distinguished from malignant tumors, such as lung carcinoma. Lung carcinoma often manifests as solid tumor mass in lung, and tumor marker level is positive. Furthermore, it often is accompanied by pleural effusion and bilateral hilar and mediastinal lymphadenopathy.

4. Conclusion

To sum up, the clinical manifestations and laboratory examination of primary pulmonary angiosarcoma are all atypical, and the diagnosis of primary pulmonary angiosarcoma is mainly established pathologically. But, our case shows that CTPA is a useful radiological tool to accurately identify primary pulmonary angiosarcoma, and it provides the important basis for early diagnosis. Therefore, CTPA occupies a very important position in the diagnosis of primary pulmonary angiosarcoma, and it is of great value in improvement of patient’s prognosis.

Acknowledgments

The authors thank the staff of the Department of cardiothoracic surgeon and radiology at The First Hospital of Jiaxing for providing valuable clinical and radiological support. In the
meantime, I would like to thank all of my friends, especially my lovely wife for her encouragement and support.

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