A Case of Pulmonary Artery Sarcoma

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Case report

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Abstract

Background: pulmonary artery sarcoma is a rare mesenchymal tumor. The prognosis of this very infrequent disease remains poor, and surgical resection is important for obtaining the best outcome.

Case Presentation: A 51-year-old male patient presented to our clinic due to the finding of the isolated lung mass in the left lower lobe of the lung. Preoperative pulmonary artery computed tomographic angiography scans showed a mass with the in the left pulmonary artery and an isolated mass in the left dorsal segment of the lung (RS6). Under extracorporeal circulation the left pneumonectomy was performed, the surgical resection margins were confirmed to be tumor-free on frozen section and all lymph nodes were free of tumor. Histopathology, immunohistochemistry, as well as fluorescence in situ hybridization (FISH) indicated the diagnosis of pleomorphic rhabdomyosarcoma.

Conclusion: This case report describes a PAS which was successfully resected with pneumonectomy and concludes that biopsy may play a limited role in confirming the pathological type and pneumonectomy should be considered in the surgical treatment of PAS.

Background

PAS is a rare, highly malignant, mesenchymal tumor. The prognosis of this very infrequent disease remains poor, and surgical resection is important for obtaining the best outcome [1, 2]. Here we report a more rare case of PAS presents as an isolated lung mass.

Case Presentation

In March 2018, a 51-year-old male patient presented to a local hospital due to cough for a month, then pulmonary CT examination was performed and found a mass at the left hilum. In April 2018, the patient went to the oncology department of our hospital, then electronic bronchoscopy revealed neoplasm in the dorsal segment of the left lower lung, histopathologic and immunohistochemical findings indicated it in agreement with the characteristics of isolated fibroadenoma and. Finally the patient was referred to the thoracic surgery department of our hospital in May 2018. He had no special past medical and family history. Physical examination showed no abnormalities, Oxygen saturation was 97%. D-dimer was 0.48 (ug/ml). CYFRA21-1 was 1.9 (0-1.8 ng/ml) The electrocardiogram showed no ST changes. Transthoracic echocardiography showed normal left ventricular function (ejection fraction 60%), and no findings of pulmonary arterial hypertension. Pulmonary artery CTA revealed occlusion with filling defect in the left pulmonary arteries and an abnormal shadow in the hilum of the left lung. (Fig. 1a, 1b arrow designating tumor). Magnetic resonance imaging (MRI) revealed a round mass. Filling defects can be seen in the trunk and distal branches of the left pulmonary artery. PET-CT showed that nodules in the dorsal segment of the lower lobe of the left lung were more likely to be sarcomas with SUVmax 11.9, while lumps in the left pulmonary artery were tumors or tumor plugs with SUVmax 9.8.
Left pneumonectomy resection was performed in May 30, 2018. A Cardiopulmonary bypass was established by inserting venous blood through the right ventricular outflow tract into the right atrium, and the artery was cannulated through the thoracic aorta (Fig. 2a). Under cardiopulmonary bypass, the patient underwent left pneumonectomy through the fourth intercostal lateral incision. After dissection of the left mainstem bronchus, the left main bronchus was detached 1 cm from the carina and the left pulmonary artery was severed at the origin of the left pulmonary artery and repaired with an autologous pericardium. The proximal resection margins of the left pulmonary artery were confirmed to be tumor-free on frozen section. The left lung was removed (Fig. 2b, c arrow designating tumor). At the same time, lymph nodes L7, L9 and L10 were biopsied. Studies of histopathology, immunohistochemistry, as well as fluorescence in situ hybridization (FISH) indicated the diagnosis of pleomorphic rhabdomyosarcoma (Fig. 2d). The tumor stage is T4N0M0. The patient was discharged on the 9th day after the operation in a satisfactory condition. He was followed up by telephone interview. The patients received chemotherapy after the operation, but we could not get the specific chemotherapy regimen of the patient. He remains alive on follow-up 18 months after diagnosis.

**Discussion And Conclusion**

PAS is very rare and highly malignant neoplasm with an aggressive behavior with an incidence of 0.001–0.03% among autopsy cases and making up roughly 15% of all primary cardiac tumors [3, 4]. Less than 400 PAS cases have been reported since it was described by Mandelstamm in 1923. It generally derives from the endothelial cells of the pulmonary artery, and tend to spread within the pulmonary vasculature. Rarely, the pulmonary parenchyma itself is invaded (usually only in the later stages), and, unlike angiosarcomas elsewhere [5]. In general, PAS is usually found in Middle-aged patients [6–8]. PAS is likely to be mistaken for chronic pulmonary thromboembolism because the clinical manifestations of PAS are remarkably similar to those of pulmonary thromboembolic disease [4, 6, 7, 9, 10]. The histologic diagnosis can be determined by EBUS-TBNA and Endovascular Catheter Biopsy [11, 12]. However, in our case, the pathologic type of the biopsy was not consistent with the pathologic type after surgery. Surgical remains the mainstay of treatment for PAS since complete resection could offer the only chance for a potential cure. Surgical options range from radical resection, such as lobectomy and pneumonectomy, to endarterectomy [13]. The median survival time has been reported between 1.5 and 12 months [14], and more recent cases have shown the benefit of improved management by aggressive surgical resection and adjuvant chemoradiotherapy with longer survival times reported [15]. Early diagnosis and complete surgical resection is crucial for prolonged survival. There is no widely accepted treatment protocols to guide the management of this orphan disease. In our case the tumor seemed to be controlled by surgery and adjuvant chemotherapy. There are many varieties of histopathologic patterns; thus more case reports are warranted to determine against which histopathologic types adjuvant therapy is effective and what is the most beneficial adjuvant therapy protocol.

In conclusion, biopsy may play a limited role in confirming the pathological type, aggressive resection of PAS is beneficial to improve the prognosis of patients, adjuvant chemoradiotherapy is also worth undertaking to improve the prognosis.
Abbreviations

PAS: pulmonary artery sarcoma

Declarations

Ethics approval and consent to participate

This study was approved by the Ethics Committee of Second Xiangya Hospital of 173 Central South University, Changsha, China. Written informed consent was obtained from the patient reported in this study.

Consent for publication

Written informed consent for publication of the clinical details and/or clinical images was provided by the patient.

Availability of data and materials

As a case report, all data generated or analyzed are included in this article.

Competing interests

The authors have no conflicts of interest to declare in this work.

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Authors’ contributions

YKL drafted and edited this manuscript and analyzed the patient data. YH edited this manuscript and analyzed the patient data. WH, FW, JQT, QCL, and FLY analyzed the patient data. WH prepared and assessed the histopathological images. WLL performed the surgery, edited this manuscript, and analyzed the patient data.

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