Case Report

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Pulmonary mucormycosis: a case of pulmonary arterial hypertension, Westermark sign, and bronchopleural fistula

Weipeng Shao[1], Zhenrong Zhang[2], Hongxiang Feng[2], Chaoyang Liang[2] and Deruo Liu[1]

Abstract

We herein describe a patient with pulmonary mucormycosis and acute myelogenous leukemia. Computed tomography showed a widened pulmonary artery, a bronchopleural fistula, and the Westermark sign. Despite worsening hemoptysis, the operation was delayed for 6 months. The operation was very complicated and difficult. A thorough preoperative examination, adequate preoperative preparation, appropriate surgical timing, and rich clinical and surgical experience were the keys to successful surgery in this case.

Keywords

Pulmonary mucormycosis, bronchopleural fistula, pulmonary arterial hypertension, Westermark sign, surgery, case report

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Introduction

Pulmonary mucormycosis (PM) is a relatively rare but fatal infection that mostly occurs in immunocompromised persons.[1] Diabetes, malignancy, and solid organ transplantation are the most common underlying conditions.[2] PM is an infection caused by the inhalation of spores, which results in pneumonia with subsequent

[1]Department of General Thoracic Surgery, Peking University China-Japan Friendship School of Clinical Medicine and China-Japan Friendship Hospital, Beijing, China
[2]Department of General Thoracic Surgery, China-Japan Friendship Hospital, Beijing, China

Corresponding author:
Deruo Liu, Department of General Thoracic Surgery, Peking University China-Japan Friendship School of Clinical Medicine and China-Japan Friendship Hospital, No. 2 Yinghua East Road, Beijing 100029, China.
Email: deruoliu@163.com

necrosis and infarction of lung tissue.[3] The infection can also spread directly or hematogenously to surrounding structures in the mediastinum, such as the heart or other organs.[4] Although the management and outcomes of PM have improved during the past few decades, the mortality rate remains high. Mortality varies with the site of infection: in previous studies, 96% of patients with disseminated disease died and 76% with pulmonary infection died.[2,5] Successful treatment of PM requires rapid diagnosis, reversal of predisposing factors, aggressive surgical excision, and antifungal therapy.[1,4]

In the present report, we describe a patient with invasive PM complicated by pulmonary arterial hypertension (PAH), the Westermark sign, and a bronchopleural fistula.

Case summary

A 56-year-old woman (height, 150 cm; weight, 43 kg) with a history of early liver cirrhosis secondary to hepatitis C virus was diagnosed with acute myelogenous leukemia (M5b) by a bone marrow smear. Bone marrow depression developed during seven periods of induction chemotherapy. However, she achieved complete remission after chemotherapy according to a follow-up survey. She had experienced coughing with expectoration at the beginning of the disease course. Figure 1 shows the changes observed on chest radiographs (exudation, consolidation, and cavity formation) during preoperative treatment from 1 March 2019 to 16 December 2019. Additionally, chest computed tomography (CT) revealed an obviously widened pulmonary artery, a thick-walled cavity with a septum in the left upper lung lobe, consolidation in the left lower lung lobe, and left pleural effusion. Bronchoscopy showed congestion and edema of the left main bronchus, obvious stenosis at the opening of the upper and lower lobes, and a large amount of necrotic

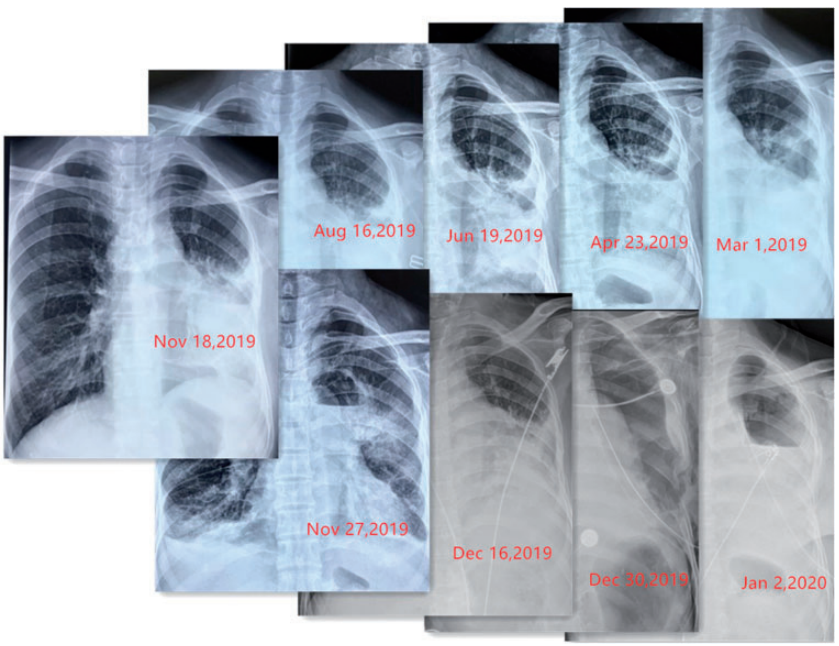

Figure 1. Changes in chest radiographs during treatment from 1 March 2019 to 16 December 2019 and after surgery from 30 December 2019 to 2 January 2020.

material attached to the bronchial mucosa. A bronchial biopsy yielded fungal organisms consistent with mucormycosis with necrosis. Given the confirmation of mucormycosis, amphotericin B (25 mg/day) and posaconazole (10 mL twice daily) were immediately initiated. Three months later, she developed hemoptysis that gradually worsened, and she therefore received hemostatic therapy (pituitrin, sodium carbon sulfonate, and hemagglutinase). The patient underwent repeated CT re-examinations. Considering the effectiveness of the hemostatic treatment and the limitations of left pneumonectomy, the patient and her family members refused surgical treatment. However, with the increasing amount and frequency of hemoptysis, the patient returned to the hospital hoping for surgical treatment. Enhanced CT revealed main pulmonary artery dilatation with an internal diameter of about 3.7 cm, and the cavity was obviously enlarged (Figure 2(a)). Echocardiographic assessment showed that the main pulmonary artery was widened (diameter of 36 mm) and that PAH was present (40 mmHg). The patient underwent right cardiac catheterization to further assess the effect of pulmonary artery widening on surgery. The maximum pulmonary artery pressure was 29 mmHg, the lowest pulmonary artery pressure was 7 mmHg, and the mean pulmonary artery pressure was 10 mmHg. The pulmonary arterial wedge pressure was 4 mmHg. The Westermark sign was also present, indicating abrupt cutoff of pulmonary vascularity distal to a large central pulmonary embolus. The presumed mechanism underlying the occurrence of this imaging sign is nearly complete obstruction of blood flow to the pulmonary artery distal to the embolic clot.

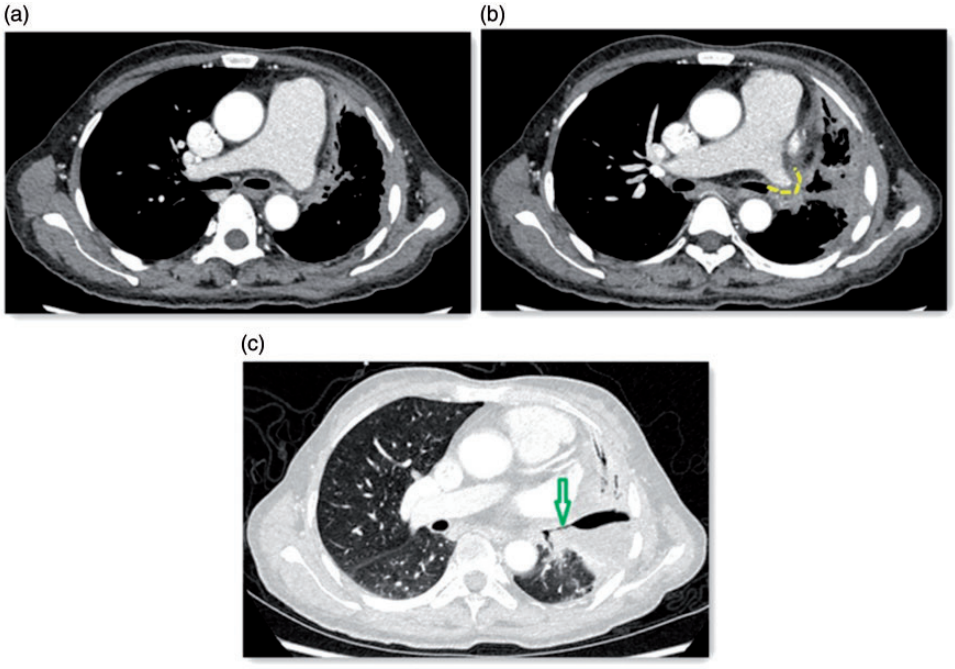

Figure 2. Computed tomographic scan of the chest showing (a) pulmonary artery dilatation with an internal diameter of about 3.7 cm, (b) the Westermark sign (dotted yellow line), and (c) a bronchopleural fistula.

The lack of flow to these more distal vessels presumably results in their radiographic transparency and the appearance of an abrupt truncation, as shown in the present case (Figure 2(b)). CT showed that the cavity communicated with the bronchus, forming a bronchopleural fistula (Figure 2 (c)). Pulmonary function tests demonstrated that the forced expiratory volume in 1 s (FEV1) was 1.35 L, FEV1/forced vital capacity was 79.04%, FEV1 percentage was 70%, and predicted diffusing capacity of the lung for carbon monoxide was 35%. After a multidisciplinary consultation involving the thoracic surgery department, respiratory department, and anesthesiology department, the patient underwent preoperative preparation followed by left pneumonectomy. Figure 1 shows the chest radiograph changes during postoperative treatment from 30 December 2019 (day of operation) to 2 January 2020.

Several difficulties were encountered during the operation. First, severe pleural adhesions were present (Figure 3(a) and (b)), causing a large amount of bleeding. Second, the thick-walled cavity on CT was a lung abscess with a diameter of about 6 cm, and a bronchopleural fistula was found. Third, the left pulmonary artery trunk was adhered to the surrounding tissue. After cutting, we found that the pulmonary artery had been completely occluded (Figure 3(c)). Fourth, the superior and inferior pulmonary veins shared a common trunk. Only the pericardium was opened for

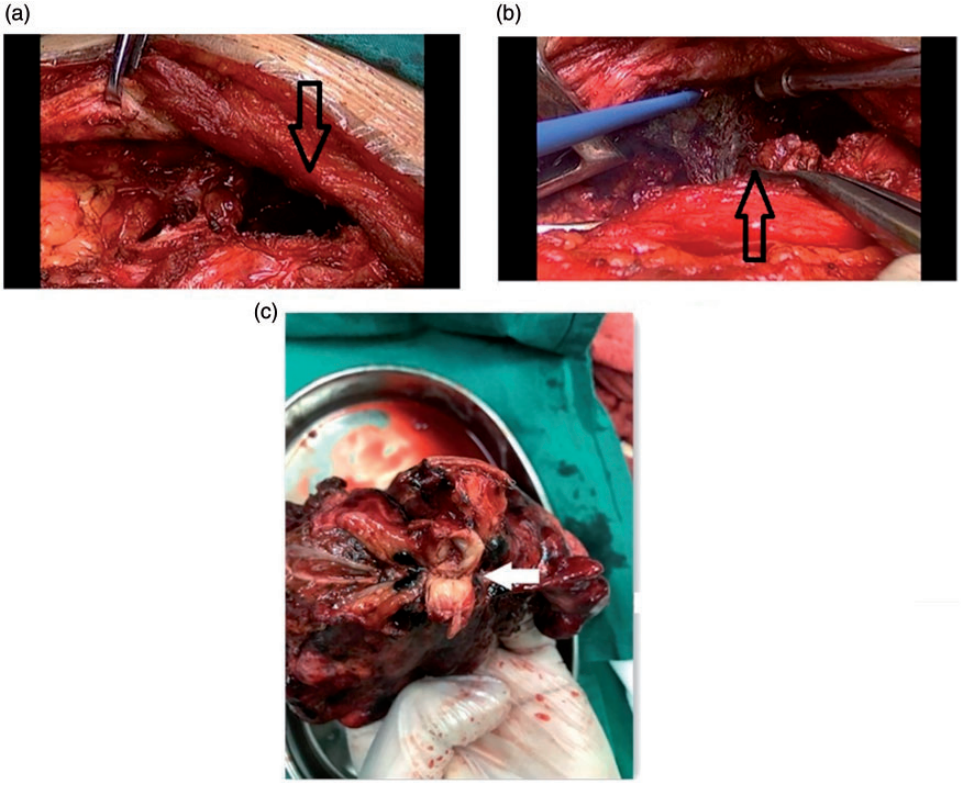

Figure 3. Intraoperative findings. (a, b) Thick-walled cavity and severe pleural adhesions. (c) Pulmonary artery occlusion.

surgery. The operation time was 230 minutes. The bleeding volume was approximately 1000 mL, and an 800-mL blood transfusion was administered. After the operation, the patient was transferred to the intensive care unit for further treatment. She was then transferred out of the intensive care unit on the first postoperative day, and the drainage tube was pulled out on the third postoperative day. The patient continued to receive intravenous liposomal amphotericin B (400 mg once daily, 5 mg/kg) and oral posaconazole (10 mL twice daily) after surgery. The intravenous liposomal amphotericin B was discontinued on day 14 postoperatively, and the oral posaconazole (10 mL twice daily) was continued as the sole therapy. Unfortunately, the patient developed a relapse of acute myelogenous leukemia 1 month after surgery and died 3 months later.

Discussion

The high mortality observed among patients with PM may be related to a delayed diagnosis, a poor host response, and limited available therapy.[6] Many previous reports have mentioned the etiology, epidemiology, clinical manifestations, imaging manifestations, diagnosis, and treatment of PM.[7–9] Early diagnosis is important for early treatment of this life-threatening disease.[10] Unfortunately, our patient's symptoms were tolerable and the operation was therefore delayed. The operation was not performed until 10 months later, which increased the difficulty of the procedure. Surgical resection combined with antifungal therapy has been proven to be the best strategy for the treatment of PM.[9,11]

We focused on perioperative management in this case. PM invasion of blood vessels leads to blood vessel thrombosis and tissue necrosis, and death may even occur because of massive hemorrhage.[12] Our patient underwent enhanced CT, echocardiography, and right cardiac catheterization to determine the pulmonary artery pressure and ensure better perioperative management. The left lung had completely lost its function. The cause of the widened pulmonary artery and PAH was probably PM invasion of the perivascular tissue, resulting in vascular occlusion. The Westermark sign formed gradually, and the heart and lung gradually adapted to the ischemic conditions. Therefore, the PAH had little effect on the left pneumonectomy procedure. This was also demonstrated by the stable circulation throughout the operation. A bronchopleural fistula is a serious complication of PM,[2] but it produced no obvious symptoms in the present case. We speculate that the formation of the bronchopleural fistula was a chronic process. PM invaded the bronchus and caused inflammatory proliferation of the surrounding tissues, then enveloped the fistula and formed a thick-walled cavity. Intrathoracic adhesions are common in PM,[4] and our solution was to perform video-assisted thoracoscopic surgery to expand the field of view for separation. We found no serious tissue edema, so the operation was postponed. Although the inflammatory edema disappeared, the tissue adhesion increased in severity. In contrast, an early operation with severe edema would have also increased the difficulty of the operation. Overall, surgeons' experience in the treatment of PM remains limited, and further multicenter research is needed.

A thorough preoperative examination, adequate preoperative preparation, appropriate surgical timing, and rich surgical experience are the keys to successful surgery in cases such as that described in the present report.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics

This study was approved by our institutional review board. The requirement for patient consent was waived because of the retrospective nature of the study.

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ORCID iD

Weipeng Shao https://orcid.org/0000-0001-9077-6408

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