Case Report

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Angioinvasive Mucormycosis Mimicking Mass and Pulmonary Thromboembolism in a Patient with Myelodysplastic Syndrome: A Case Report

골수이형성 증후군 환자에서 발생한 종괴와 폐색전증을 모방하는 혈관침습성 점막진균증: 증례 보고

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Mucormycosis encompasses a range of fungal infections that can impact various organs. Although pulmonary mucormycosis is relatively rare, it poses a significant threat, particularly to individuals with compromised immune systems. Pulmonary mucormycosis presents with various radiological manifestations. Notably, the involvement of the angioinvasive pulmonary artery in pulmonary mucormycosis cases has seldom been documented. In this report, we showcase the radiological characteristics of angioinvasive mucormycosis, which can mimic pulmonary thromboembolism or a pulmonary artery tumor, in a patient diagnosed with myelodysplastic syndrome.

Index terms Pulmonary Mucormycosis; Angioinvasive Mucormycosis; Myelodysplastic Syndrome; Computed Tomography

INTRODUCTION

Mucormycosis represents a cluster of fungal infections caused by the members of the order Mucorales. It can affect many organs, and pulmonary mucormycosis, although rare, stands as a life-threatening opportunistic fungal infection (1). Typically, it

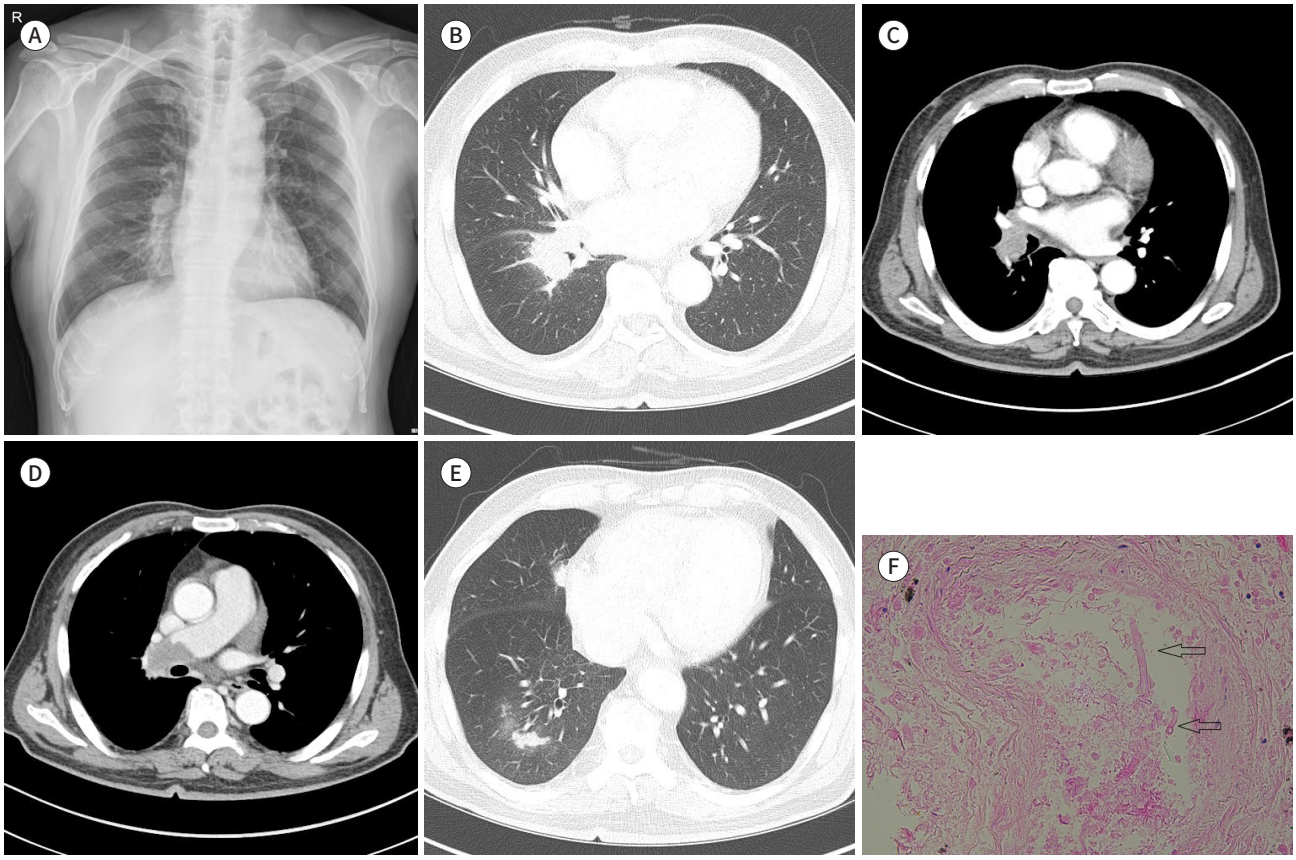
targets individuals with compromised immune systems or those harboring risk factors such as uncontrolled diabetes mellitus, hematological malignancies, solid organs, or bone marrow/stem cell transplantation (2). The incidence of pulmonary mucormycosis has been reported to be as high as 24% among mucormycosis cases, leading to elevated mortality and morbidity owing to its rapid progression and prominent angioinvasion (3, 4).

Although the radiological presentations of pulmonary mucormycosis exhibit variability, ranging from ground-glass opacity (GGO) to consolidation, the radiological characteristics of progressive pulmonary artery involvement in patients with pulmonary mucormycosis have rarely been reported. In this report, we elucidate the radiological attributes of angioinvasive mucormycosis, which can masquerade as a mass or pulmonary thromboembolism, in a patient afflicted with myelodysplastic syndrome.

CASE REPORT

A 65-year-old male patient with underlying myelodysplastic syndrome was admitted to our hospital for his scheduled third round of chemotherapy. On the 2nd day of admission, the patient had a mild fever of 37.5°C before commencing treatment, along with abnormal laboratory findings that included elevated C-reactive protein (CRP, 14.3 mg/dL, normal range: 0–0.5 mg/dL), elevated D-dimer (372 ng/mL, normal range: 0–243 ng/mL), and pancytopenia (white blood cell 310/μL, red blood cell 204000/μL, platelet 44000/μL). Chest radiography revealed a prominent right hilum with increased opacity in the central area of the right lower lung (Fig. 1A). Contrast-enhanced CT showed a mass-like consolidation in the interlobar area, associated with partial obliteration of the hilar, interlobar, and lobar branches of the pulmonary artery in the right lower lobe (Fig. 1B, C). Following the administration of empirical antibiotics, CRP levels gradually decreased until the 6th day of hospitalization. However, intermittent fever persisted, along with increased CRP levels, reaching up to 15.7 mg/dL. On a follow-up CT performed on the 8th day of hospitalization, there was a similar but slightly increased extent of the previous mass-like consolidation surrounding the bronchus intermedius and lobar bronchi of the right middle lobe and right lower lobe, displaying further subtle obliteration at the hilar, interlobar, and lobar branches of the pulmonary artery in the right lower lobe. Even with disease progression, no bacterial growth was observed in blood cultures. We considered the possibility of pulmonary thromboembolism and tumors of the pulmonary arteries, including angiosarcoma or lymphoproliferative disorders. Bronchoscopy revealed no invasive lesions in the bronchus, and histological examination of the 11R lymph nodes revealed no malignant cells. Although the possibility of pulmonary thromboembolism appeared high, considering the filling defect of the pulmonary artery and the accompanying low-grade fever, the extent of the filling defect continued to increase despite anticoagulant treatment. Consequently, we had to consider other diseases mimicking pulmonary thromboembolism. Patient condition gradually worsened, with a fever of 38.7°C and an elevation of the CRP level to 18.8 mg/dL. On chest CT performed on the 25th day of hospitalization, we observed further proximal progression of endoluminal filling defects, displaying an abrupt cut-off sign in the right hilar and main pulmonary arteries, as well as newly developed filling defects in the lobar and segmental pulmonary arteries of the right middle lobe (Fig. 1D). Mul-

Fig. 1. Progressive angioinvasive mucormycosis with direct invasion of pulmonary arteries in a 65-year-old male with underlying myelodysplastic syndrome.
A. Initial chest radiograph reveals prominent right hilar opacity with central consolidation in the right lower lung zone.
B, C. Axial lung window and mediastinal window chest CT images on the 4th day of hospitalization, depict a mass-like consolidation at the interlobar area of the right lower lobe (B). The axial narrow-window chest CT image displays mass-like consolidation obliterating the interlobar pulmonary artery in the right lower lobe (C).
D, E. Axial mediastinal and lung window and lung window chest CT images on the 25th day of hospitalization, illustrate proximal progression of the endoluminal filling defect in the right hilar and main pulmonary arteries, featuring an abrupt cut-off sign (D). Additionally, newly developed filling defects are observed in the lobar and segmental pulmonary arteries of the right middle lobe. Furthermore, the images also reveal new ill-defined nodules and patchy ground-glass opacity in the right middle and lower lobes (E).
F. Photomicrograph of the pathological specimen displays a few degenerated fungal hyphae (arrows) in the background of the necrotic lung parenchyma. These broad and aseptate hyphae correspond to Mucorales hyphae (hematoxylin & eosin stain, × 40). The final diagnosis confirmed angioinvasive mucormycosis.



tiple new ill-defined nodules and patchy GGOs in the right middle and lower lobes were also evident (Fig. 1E). Finally, we considered the possibility of an angioinvasive mucormycotic infection, showing direct invasion of the proximal pulmonary artery with pulmonary thrombi and disseminated lung nodules. A percutaneous needle biopsy was performed to target the consolidation in the right lower lobe. This revealed a few degenerated fungal hyphae in the background of the necrotic lung parenchyma, characterized as broad and aspartate, consistent with the characteristics of Mucorales. The final diagnosis confirmed definite angioinvasive mucormycosis (Fig. 1F). The patient was administered intravenous amphotericin B (350 mg/day, 5 mg/kg). Despite medical treatments, including antibiotics and antifungal agents, patient lung condition did not improve. On the final follow-up CT scan, further progression of

filling defects was observed in multiple pulmonary arteries, new septic embolic infarctions, and multiple pulmonary disseminated mucormycotic nodules. Unfortunately, the patient did not recover and passed away 5 months after admission.

This study complies with the Declaration of Helsinki and written informed consent was obtained from the patient.

DISCUSSION

In this report, we present a distinctive case of angioinvasive mucormycosis that extensively and directly affected the proximal large pulmonary artery. This presentation mimicked pulmonary thromboembolism and malignancy, with the subsequent development of multiple pulmonary nodules, indicating the spread of fungal infection to the lung parenchyma. This occurred in a patient with myelodysplastic syndrome.

Mucormycosis is a relatively rare fungal infection in humans, but when it does occur, it can lead to severe angioinvasive infections. Most patients afflicted by this invasive disease have underlying conditions, such as hematologic malignancies, that make them susceptible to infection (5). The spores of these fungi, which are commonly found in soil, fallen leaves, compost, animal dung, and the air, can be inhaled, resulting in lung and sinus infections that can extend into the brain and eyes.

Although the appearance of pulmonary mucormycosis can vary on imaging, lobar and segmental consolidation, multiple nodules, and masses with a ground-glass halo or central GGO surrounded by dense consolidation—the so-called reversed halo sign. Multilobar distribution is also frequently observed. If left untreated, unilateral infections can rapidly progress to the contralateral lung. A multifocal pneumonia pattern with bilateral consolidation is strongly associated with increased mortality (1). Cases involving direct pulmonary vascular invasion by mucormycosis may exhibit pseudoaneurysm formation or an abrupt termination of the pulmonary artery branch, which can manifest as a vascular cutoff sign (1). In some cases, small pulmonary arterial filling defects can be observed in macroscopic septic emboli (1). In this case, we identified similar findings, such as the proximal pulmonary vascular cutoff sign and mass-like consolidation, although we initially misinterpreted them. Furthermore, the predominant involvement of the proximal interlobar and hilar pulmonary arteries, along with the involvement of the main pulmonary artery and the delayed appearance of typical lung nodular lesions, added to the complexity of the diagnosis. Importantly, no evidence of pseudoaneurysm formation was found in the affected pulmonary arteries, cavitation, multifocal pneumonia, or the halo signs associated with parenchymal lung lesions (1). Furthermore, the absence of a reversed halo sign, a highly specific marker of pulmonary mucormycosis, further contributed to the diagnostic challenge (6). Few reported cases where filling defects in the pulmonary arteries grew rapidly without vessel dilatation, mimicking massive pulmonary thromboembolism or angiosarcoma, as observed in our report. In our case, we believe that angioinvasion in the interlobar area of the right lower lobe may have subsequently propagated to the proximal hilar and main pulmonary arteries, leading to dissemination in the bilateral lungs and the emergence of lung nodules. This case underscores that angioinvasive mucormycosis can present as rapidly progressing filling defects in the pulmonary arteries, leading

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to misdiagnosis as other malignant or vascular diseases.

Mucormycosis carries a grim prognosis, marked by elevated morbidity and mortality rates among affected individuals. The likelihood of higher mortality increases when there are delays in accurate diagnosis and the initiation of suitable treatment, often stemming from misdiagnosis. Consequently, it underscores the significance of recognizing the diverse imaging characteristics of angioinvasive mucormycosis, particularly in patients with hematologic diseases, to facilitate early diagnosis.

In summary, we presented a distinctive case of angioinvasive mucormycosis, which remarkably imitated pulmonary thromboembolism and a pulmonary artery tumor within the context of pulmonary mucormycosis. This occurred in a patient already grappling with myelodysplastic syndrome.

Author Contributions

Conceptualization, K.S.S.; supervision, K.S.S., C.S., K.J.H., K.H.; writing—original draft, N.H.J.; and writing—review & editing, K.S.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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골수이형성 증후군 환자에서 발생한 종괴와 폐색전증을 모방하는 혈관침습성 점막진균증: 증례 보고

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Mucormycosis는 Mucorales에 의해 유발되는 진균 감염으로, 다양한 장기를 침범할 수 있으며 그중에서도 폐점균증은 드물지만 특히 면역 저하 환자에서 생명을 위협하는 기회 감염이다. 폐점균증의 영상 소견은 다양하지만, 폐점액진균증 환자에서 점점 커지는 폐동맥색전증을 모방하는 경우는 드물게 보고되었다. 저자들은 골수이형성 증후군 환자에서 종괴와 폐색전증을 모방한 혈관 침습성 점막 진균증의 증례에 대해 보고하고자 한다.

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