

Successful Treatment of Concurrent Mucormycosis and Diffuse Alveolar Hemorrhage in a Patient With Newly Diagnosed Systemic Lupus Erythematosus

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Infection is often complicated with systemic lupus erythematosus (SLE) and is one of the leading causes of death in SLE patients.¹ When SLE flare coexisting with serious infection is suspected, intensive immunosuppressive therapy is usually a challenge. Mucormycosis is a rare and rapidly progressing infection with high mortality. Diffuse alveolar hemorrhage (DAH) is also one of the rare and serious manifestations in SLE.^{2,3} We experienced an extremely rare case of SLE that was initially presented with DAH and mucormycosis concurrently.

A previously healthy, 42-year-old female patient was admitted with a one-month history of loss of appetite, weight loss, and rash on her face and extremities.

Complete blood count revealed leukopenia and mild anemia. Urinalysis showed microscopic hematuria and proteinuria. Both erythrocyte sedimentation rate and C-reactive protein were slightly elevated. Immunologic studies were positive for antinuclear (homogeneous pattern, 1:1280), anti-Ro, anti-Sm, and anti-ribonucleoprotein antibodies, but anti-neutrophil cytoplasmic antibody was negative. Anti-double stranded deoxyribonucleic acid was elevated at 62.3 IU/mL and complements were low. Lupus anticoagulant and direct Coombs' test were

positive. Abdominopelvic computed tomography (CT) showed multiple enlarged lymph nodes and borderline splenomegaly. Biopsies from skin lesions, right inguinal lymph nodes, and bone marrow were performed and their histopathology revealed leukocytoclastic vasculitis, reactive hyperplasia, and normal findings, respectively.

On the fifth day of hospitalization, she complained of cough and shortness of breath, and chest radiography showed consolidations in bilateral lower lung fields. Intravenous antibiotics were started for a presumed pulmonary infection. No microorganism was grown in the sputum, blood or urine, and further microbiologic tests were all negative for cytomegalovirus, aspergillus and *Pneumocystis jirovecii*. High-dose glucocorticoid was added for suspected concurrent lung involvement in newly developed SLE, but symptoms and lung infiltrations on CT (Figure 1a) worsened and black-colored crusted patches occurred on the right ala of her nose (Figure 2a). On the 17th hospital day, the patient was transferred to the intensive care unit (ICU) and placed on mechanical ventilator due to expected impending respiratory failure. A bronchoscopic exam showed bloody bronchoalveolar lavage fluid. Biopsy of the nasal lesion and histopathology confirmed mucormycosis (Figure 2b). Plasma

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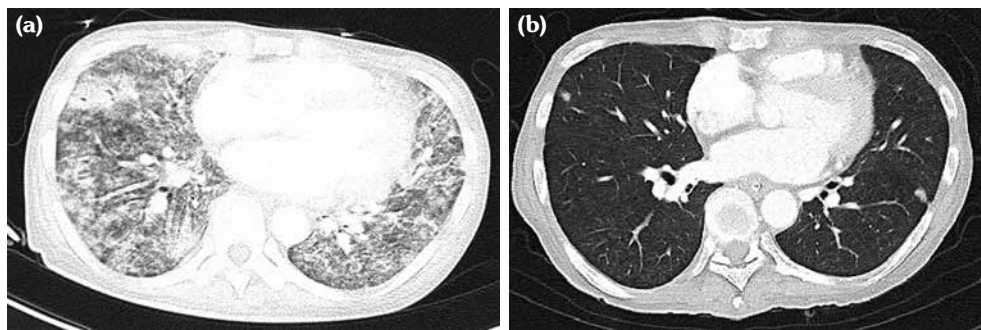


Figure 1. (a) Chest computed tomographs showed bilateral, diffuse, ill-defined, nodular consolidation and ground-glass opacification in lungs, suggesting diffuse alveolar hemorrhage. (b) The lung lesions were markedly resolved after cyclophosphamide and glucocorticoid pulse therapies.

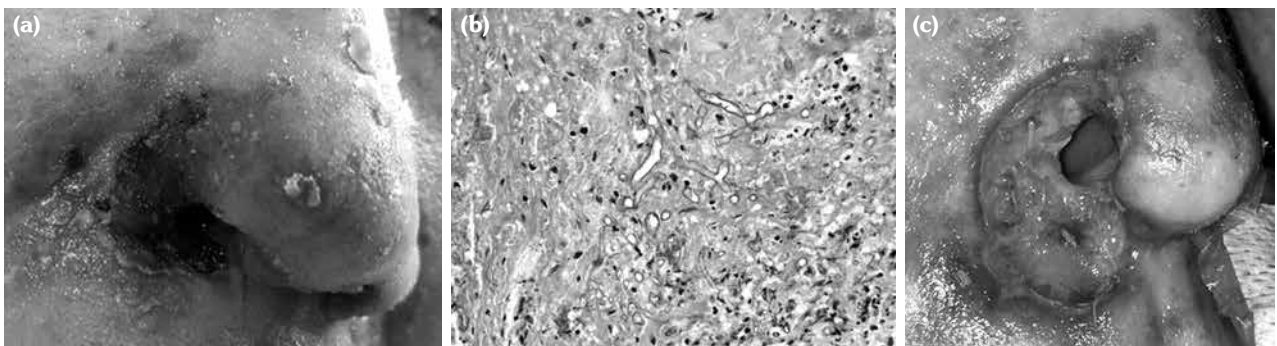


Figure 2. (a) Black-colored crusted patches on the right ala of nose were confirmed to be mucormycosis. (b) Periodic acid schiff staining of excised nose tissue showed thick fungal hyphae with right angle branching (magnification 400 \times) (c) Mucormycosis infected tissue was removed by surgical debridement.

exchange and intravenous immunoglobulin therapy were performed with liposomal amphotericin B. However, her respiration was not improved and the endotracheal tube was frequently blocked because of persistent DAH. Notwithstanding the risk of the concurrent infection aggravation, cyclophosphamide and glucocorticoid pulse therapy were added for the management of the refractory DAH. Three cycles of cyclophosphamide pulse therapies (intravenous cyclophosphamide 500 mg with methylprednisolone 1 g) were administered with a surgical debridement of the nasal mucormycosis lesions on the 39th hospital day (Figure 2c). Her respiration stabilized and the lung lesion gradually resolved with the cyclophosphamide pulse therapies (Figure 1b). The patient eventually recovered after three months of ICU care.

Systemic lupus erythematosus patients with flare have an increased risk of infection.^{4,5} Thus,

controlling SLE disease activity may be important for infection management. This case suggests that the prompt start of intensive immunosuppressive therapy with aggressive management of infection may result in a favorable outcome in SLE patients with a concurrence of flare and serious infection.

Declaration of conflicting interests

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