Necrotizing Sarcoid Granulomatosis: A Case with Severe Progression

To the Editor,

A 40-year-old man presented with a 10-year history of progressive dyspnea, recurrent hemoptysis, and fatigue. In 2004, he had been diagnosed with sarcoidosis based on an open lung biopsy. He refused any treatment and had irregular outpatient monitoring. He was an active smoker with no other comorbidities. His physical examination was unremarkable; oxygen saturation rate measured with pulse oximetry was 95% at admission. Retrospective analysis of his imaging studies revealed convergent, cavitating nodules in central areas (Figure 1). On his last chest computed tomography image, there were multiple cavities with air-fluid levels of a maximum diameter of 7.5 cm at left upper lobe and 9 cm at right upper lobe, and tree-in-bud pattern scattered through both lungs. There were no evident lymphadenopathies. Antinuclear antibody and anti-neutrophil cytoplasmic antibody test results were negative, angiotensin converting enzyme level was within normal limits, and serum galactomannan level was 1.71 (0-0.5 index). Sputum culture, acid-fast bacilli staining, and tuberculosis culture were all negative. Previous pathology specimens were reevaluated and the histology revealed multiple giant cell formations associated with several sarcoid-like granulomas in damaged lung parenchyma, and extended ischemic necrosis and granulomas in the vessel walls. The patient was diagnosed with necrotizing sarcoid granulomatosis (NSG) and aspergilloma. NSG is one of the 5 syndromes of pulmonary angiitis and granulomatosis described by Liebow in 1973 (1). Radiological features may involve alveolar infiltrates, nodular lesions with or without cavitation, and bilateral lymphadenopathies (2). Histologically, the disease is characterized by sarcoid-like granulomas, granulomatous vasculitis involving the walls of muscular arteries and veins, and varying degrees of necrosis (3,4). The disease has a benign prognosis, and treatment consists of corticosteroids and immunosuppressive agents (5). This case demonstrates the severe destruction of pulmonary parenchyma caused by necrotizing sarcoid granulomatosis when the disease is misdiagnosed and not treated. Systemic corticosteroids and antifungal therapy was planned for the patient but unfortunately, he was again lost to follow-up.

Figure 1: Thorax computed tomography images show centrally located nodular lesions in the earliest scans and their progressive cavitation over time. In images from 2011, there is also central opacification of cavities. An intercavitary lesion suggestive of a fungus ball is noted in the left lung in the right upper image.
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CONFLICTS OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
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