

Necrotizing sarcoid granulomatosis: A case report

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Abstract

We report a case of a 22-year-old man with persistent cough and sarcoidosis-like changes in computed tomography scan. An extensive differential diagnosis is discussed and its evolution and treatment is presented.

Introduction

Necrotizing sarcoid granulomatosis (NSG), first described in 1973, is a rare systemic disease, characterized by granulomas accompanied by variable amounts of necrosis and vasculitis with no evidence to support an infectious etiology.¹

Case Report

A 22-year-old male college student was admitted to our hospital due to productive cough, fatigue, sweating, dyspnea and bloody phlegm. He was treated with amoxicillin and clavulanic acid, but maintained dry cough and night sweats. Mantoux tuberculin skin test and microbiological examination of bronchial secretions were negative for tuberculosis. There was no history of recent travels, no drug abuse and no sick cohabitants. No history of recurrent infections in childhood. He was an active smoker (1 pack-year) and had no other personal background. Physical evaluation, blood tests (except for a high sedimentation rate (22 mm, normal range: 0-10) and respiratory function tests were normal. Cultural and serological studies ruled out most of the possible infectious diagnoses: *Streptococcus pneumoniae* urinary antigen, *Legionella pneumophila* urinary antigen, *Chlamydia pneumoniae*, *Mycoplasma pneumoniae*, *L. pneumophila* serology, *Rickettsia*

conori serology, *Aspergillus fumigatus* serology, *Brucella*, *Bartonella*, *Legionella*, *Toxoplasma gondii* serology, *Echinococcus* serology, *Schistosoma* serology.

A computed tomography (CT) scan revealed enlarged lymph nodes in the mediastinum and pulmonary hila, and multiple peribronchovascular irregular consolidations with air bronchogram, throughout both lungs, with predominance in the upper and middle lung zones. Furthermore, homogeneous hepato-splenomegaly (18 and 15 cm diameter, respectively) and enlarged retroperitoneal lymph nodes were observed (Figure 1). Immunophenotypic analysis of peripheral blood, needle aspiration biopsy of lymph node and bronchoalveolar lavage were negative for lymphoproliferative disease. Selective immunoglobulin A deficiency (<5 mg/dL) and IgG4 (1.6 mg/dL; normal range: 7-89) was identified. Screening tests for autoimmune disease [rheumatoid factor and anti-ENA (antibodies to extractable nuclear antigens: anti-RNP, Anti-Sm, Anti-SSA, Anti-SSB), anti-double stranded DNA, Antinuclear antibodies, Antimitochondrial antibodies, smooth muscle antibodies, anti-neutrophil cytoplasmic antibodies] were negative. Echocardiogram, testicular ultrasound and eye examination were unremarkable. Flexible bronchoscopy showed diffuse inflammatory signs and increased resistance throughout the bronchial tree. Conventional transbronchial needle aspiration biopsy was performed on the lymph nodes which was negative for malignant cell. Endobronchial brush (performed 3 times) was negative for malignant cell as well as acid-fast bacilli.

In order to clarify the diagnosis, surgical biopsy was considered. Microscopic analysis from mediastinum lymph node, obtained by mediastinoscopy, demonstrated necrotizing granulomatous inflammation and excisional biopsy of a cervical lymph node showed necrotizing granulomatous lymphadenitis. Microbiology studies did not identify bacteria, mycobacteria or fungi. Serum angiotensin converting enzyme, serum and urinary calcium were normal.

A CT scan performed 3 months later demonstrated massive lymphadenopathy in the mediastinum, from the supra clavicular regions to subcarinal space. These lymphadenopathies were more hypodense, suggesting intra lesion necrosis. In the lung parenchyma, the peribronchovascular consolidations were similar to the previous examination. However, a perilymphatic micronodular pattern was now observed along with nodular thickening of the bronchovascular bundles, with upper and middle lung zones predominance. Also, hepato-splenomegaly and enlarged retroperitoneal

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nodes, with no significant change from the previous study (Figure 1). A positron emission tomography scan revealed intense 18F-fluorodeoxyglucose uptake in multiple bilateral pulmonary densification and bilateral supraclavicular nodes, para-tracheal, subcarinal, hilar, peri-esophageal and near the gastroesophageal junction, peri-gastric, hepatic hilum, peri-pancreatic, peri-splenic, mesenteric, inferior vena cava and right common iliac artery.

He started treatment with prednisolone (0.5 mg/Kg/day) maintaining the same dosage for 3 months. Progressive tapering was made for 6 months up to 15 mg per day. The patient stopped the medication on his own initiative at this point. There was complete resolution of symptoms and persistently normal inflammatory markers on the first month. The imaging evolution is illustrated in Figure 1, with resolution of lymphadenopathies and almost complete resolution of the lung consolidations. He maintains follow-up and is asymptomatic to date.

Discussion and Conclusions

NSG is a distinct rare pulmonary granulomatous disease of unknown etiology that

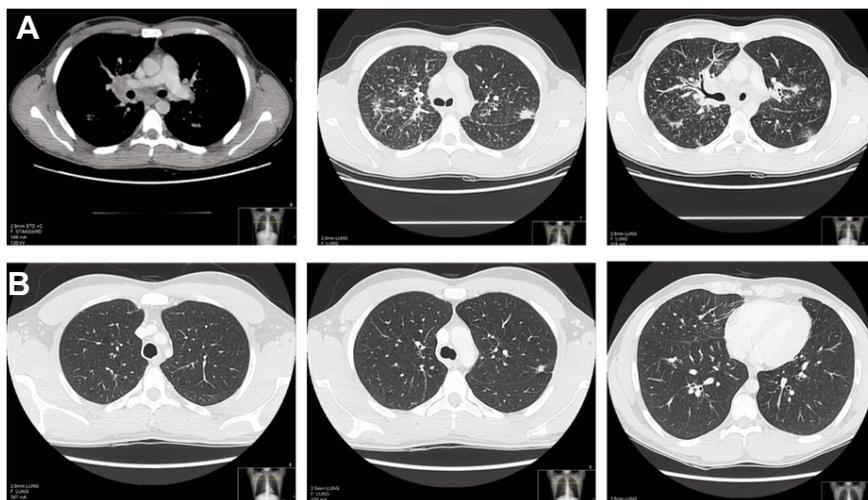


Figure 1. Thoracic computed tomography (CT) scan (A) shows multiple peri-bronchovascular consolidations, with air bronchogram and a perilymphatic nodular pattern in the upper and middle lung zones, and mediastinal and hilar lymphadenopathies. 2 years follow up CT (B), after treatment, shows no lymphadenopathies in the hila or mediastinum, and almost resolution of the pulmonary abnormalities (below).

appears to be an intermediate between sarcoidosis and systemic necrotizing vasculitides. It is characterized by sarcoid-like granulomas along with extensive necrosis and vasculitis following a usually benign clinical course.² Differential diagnoses include tuberculosis, Wegener's granulomatosis, lymphomatoid granulomatosis, neoplasms, hypersensitivity pneumonitis or Churg-Strauss syndrome. The major chal-

lenge is to ensure that it is not an infectious disease, since the treatment is based on corticosteroids. Three main parenchymal CT features are typically reported: diffuse infiltrates, bilateral perilymphatic nodules and solitary nodules.³ The prognosis of NSG patients was described as being favorable in previous reports, but fatal complications have been reported.

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