



(CASE REPORT)



Necrotising sarcoid granulomatosis: A report case

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Abstract

Sarcoidosis is a systemic granulomatous disease characterised by the appearance of noncaseifying epithelioid granulomas in the affected organs, most commonly the lungs, skin, and eyes. Necrotizing sarcoid granulomatosis is challenging to diagnose due to its rarity and similarity with other necrotizing disorders. We describe a case of a 47-year male with fever, chest pain, and nodular pulmonary opacities on chest radiograph. Histological examination of the lung tissue confirmed NSG. Diagnostically, infectious causes, vasculitis, and malignancy were excluded. A corticosteroid treatment has been started with a tendency to regression. The key to diagnosis, emphasized in our paper, is the histopathological finding. The course of NSG is similar to sarcoidosis. The treatment of choice is Corticosteroids, but the disease exhibits a tendency towards spontaneous regression.

Keywords: Necrotizing; Sarcoid; Granulomatosis; Caseous necrosis; rare entity; Differentiation diagnosis

1. Introduction

Since its first description in 1973 by Liebow (1), necrotising sarcoid granulomatosis (NSG) is a rare systemic disease, characterised by sarcoid-like granuloma, vasculitis and variable degrees of necrosis. several additional series and a few case reports have been published. This uncommon entity's diagnostic criteria include exclusion of other disorders with necrotizing granulomas, mainly GPA, rheumatoid nodule, tuberculosis (TB), and parasitic and fungal infections (2). Here we discuss a case of NSG in a 47-year-old male who had an incidental finding of hilar, mediastinal, and abdominal lymphadenopathy on imaging. Consequently, the focus is to highlight how sarcoidosis, in addition to being a multisystem disorder, can also manifest as rare histological features. This knowledge of rare variants of this disease process can help make a timely diagnosis and give high-value care to the patients.

2. Case report

A 47-year-old man, non-smoker, with no particular personal history, his mother was treated for tuberculosis in 1986 and his sister was being followed for a rheumatic disease. 3 months prior to admission, the patient presented with right basithoracic stabbing pains with no other systemic signs, all evolving in a context of apyrexia and weight loss of 13kg. The physical examination was unremarkable.

The chest X-ray showed bilateral nodular opacities with a left hilar opacity. Laboratory tests on admission showed: lymphopenia 1130 ele, no eosinophilia. C-reactive protein (CRP) 1.10 mg/l (0-5), angiotensin-converting enzyme (ACE) 109 U/l (20-70). Serum calcium 94mg/l, serum phosphorus, 24-hour calciuria and 24-hour phosphaturia were normal. Serum protein electrophoresis showed polyclonal hypergammaglobulinemia. The immunological screening including ANCA (antineutrophil cytoplasmatic antibodies) was negative. Viral serologies (HIV, HCV, HBsAg, HBcAb, HBsAb) were

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negative. Liver and kidney function tests were normal. Sputum tests for bk and gene xpert were negative, with negative quantiferon.

A high-resolution CT scan showed dense intraparenchymal and subpleural pulmonary nodules with regular contours in the upper lobar, ventral culmen, lingular, left ventro-basal and middle lobar regions, the largest measuring 10 mm long. Diffuse intraparenchymal and subpleural micronodules in both lungs, predominantly in the ventral and middle lobar, dense, regular in outline and predominantly perilymphatic in distribution. Focuses of pulmonary micronodules of bronchiolar distribution, some of which are the site of calcifications. Bilateral peribronchovascular micronodular thickening of the hilar regions. Diffuse thickening of the septal lines with a beaded scissure appearance. Bilateral thickening of the pleural cap. Mediastinal nodes. An abdomino-pelvic CT scan was performed, showing a lack of opacification with evidence of an endoluminal thrombus extending to the iliac veins. Associated with endoluminal thrombus of the portal vein and superior mesenteric vein. Transversely elongated liver of increased size homogeneous in size and regular in outline.

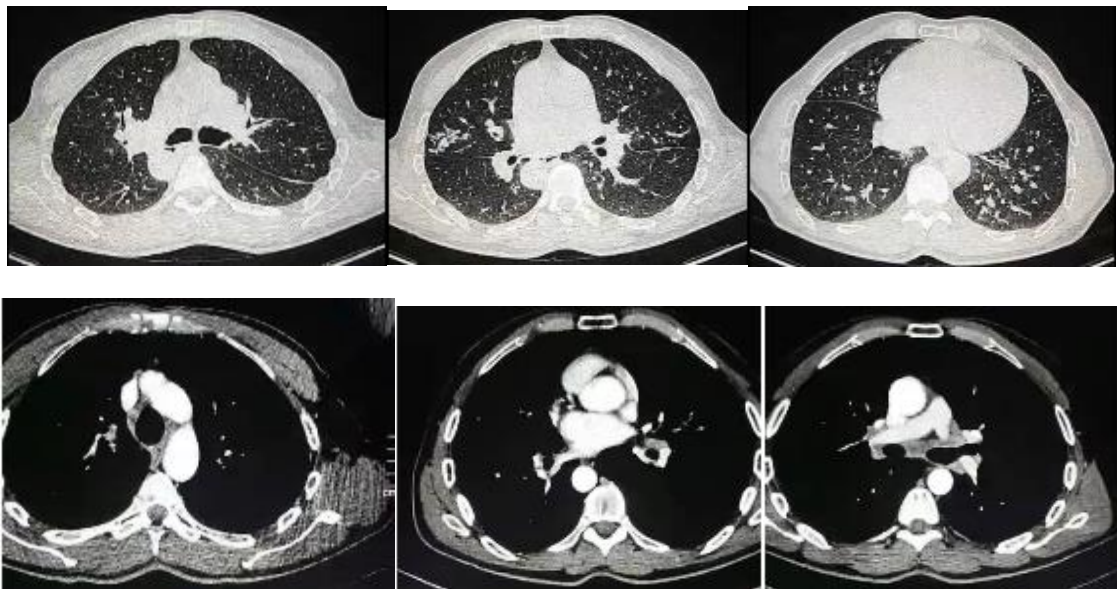


Figure 1 A follow-up contrast-enhanced chest CT performed at our hospital. (A—C) On lung window setting, intraparenchymal and subpleural pulmonary nodules. predominantly in the ventral and middle lobar Peribronchovascular distribution of the pulmonary nodules is well demonstrated (arrows). (E,F) On mediastinal window setting, Mediastinal nodes with calcifications

Bronchoscopy showed a 1st degree inflammatory state with a few granulations at the entrance to the right main bronchus and diffuse anthracotic spots. Staged bronchial biopsies: multiple granulomatous EGC lesions with the beginnings of caseous necrosis. Bronchoalveolar lavage showed predominantly lymphohistiocytic alveolitis, with no specific character.

Aspirations for cytodiagnosis: no sign of malignancy, Ziehl–Neelsen staining and polymerase chain reaction of *Mycobacterium tuberculosis* using the obtained specimen were negative.

Salivary gland biopsy revealed discrete, non-specific interstitial sialadenitis of Chisholm and Mason grade 1.

Pulmonary function tests revealed 170% RV/CPT dynamic hyperinflation; no diffusion disorder. Arterial blood gas analysis was also normal. Cardiac ultrasound was normal.

Corticosteroid therapy based on prednisone 40 mg/day was started, with curative dose anticoagulant treatment based on rivaroxaban 15 mg*2 for 21 days, then repeated at 20mg/day. Dyspnea and respiratory function abnormalities subsided after one month, and radiographic abnormalities began to resolve.

3. Discussion

Sarcoidosis is a disease that usually involves various systems of the body. Around 90% of patients who present with sarcoidosis have significant lung manifestations, including but not limited to hilar lymphadenopathy, reticular opacities, and parenchymal nodes on imaging (3). Sarcoidosis can cause epithelioid cell granuloma with marked necrosis that is generally noted in infectious disease such as mycobacterial and fungal infections. So, the correlation of pathological findings with clinical, microbiological or radiological data is necessary to determine an etiology. Because the treatment for each disease is completely different, it is necessary to proceed with great care. Granulomas are characterized as necrotizing or non-necrotizing based on the presence or absence of necrosis (4).

Necrotizing sarcoid granulomatosis was first described in 1973 by Liebow, who noted the histological presence of confluent epithelioid granulomas with small central necrosis foci or more extensive necrosis, as well as vasculitis (1). The most important differentiation is an infectious etiology such as Mycobacterium or fungi when there is necrosis in granulomas, because infection frequently causes granulomas with necrosis (2,5). Liebow differentiated this granulomatous disease from other forms of noninfectious pulmonary angiitis and granulomatosis: Wegener's granulomatosis, Churg–Strauss syndrome, bronchocentric granulomatosis, and lymphomatoid granulomatosis. Actually most authors esteem the entity as a form of sarcoidosis more than a distinct entity, differing in the fact that there is more intense necrosis and vasculitis (6). Sarcoidosis with NSG pattern can be easily misdiagnosed as pulmonary malignancy or other pulmonary granulomatous diseases due to their similar radiologic findings and nonspecific clinical presentation. Sarcoidosis with NSG pattern has been reported in a wide range of age groups from 8 to 68 years, with a median age of 42 years.[7] It occurs more constantly in women than in men with variable prevalence of extrapulmonary involvement such as eyes, skin, and nerve systems.[7] Karpathiou et al have reviewed clinical, radiologic, and histopathologic findings of NSG and found that the pattern of multiple lung nodules is the most common radiologic presentation, followed by a solitary nodule or mass. Thoracic lymphadenopathy and cavitation of nodules are often observed. Therefore, it is required to distinguish radiologic features of NSG from those of malignancy (7).

The patient described in our case study had aspects suggestive of both classic and necrotizing sarcoidosis: an elevated ACE level, intrathoracic lymphadenopathy, and necrosis within granulomas on histology. Sarcoidosis has traditionally been related with a non-necrotizing histological pattern; our case study shows that necrosis on histology might not necessarily rule out the diagnosis. NSG should be kept in the differential of necrotizing granulomatous process. Lymphoma might commonly be confused with NSG due to an extensive lymph node involvement. Hence, NSG's definite diagnosis requires excluding a malignant, infectious, or autoimmune process that might present with granulomas (8).

The most commonly used treatment for sarcoidosis with NSG pattern is systemic steroid therapy (daily prednisone 40–60 mg for 4–8 weeks).[9] Unlike other systemic vasculitis or malignancy, good response with systemic corticosteroid treatment is expected. The disease also can show spontaneous regression.[10] This was well demonstrated in our case. Overall prognosis of sarcoidosis with NSG pattern is favorable,[10] which is a notable distinguishing feature in comparison with other differential disease entities. Therefore, minimally invasive diagnostic procedure can be considered for better management of the patient.

4. Conclusion

Recognizing and understanding the clinical and radiologic presentations of sarcoidosis with NSG pattern can lead to appropriate diagnosis and management. In addition, given the relative benign nature of necrotising sarcoid granulomatosis, bronchoscopy should be performed to rule out cancer or infection. Further large case studies are needed to specify and better define this rare disease entity.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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