

Nodular Pulmonary Sarcoidosis

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SARCOIDOSIS IS A MULTIORGAN DISEASE THAT

most commonly affects adults in the twenty to forty year age group. Typically symptoms are referable to the lungs and a chest radiograph is obtained. The radiographic findings are often so typical that the diagnosis can almost be certain without further investigation. The radiographic findings characteristic of sarcoidosis are bilateral symmetrical hilar lymphadenopathy, often accompanied by right paratracheal nodes with or without parenchymal air space disease. High resolution CT scan typically shows micronodules with a perilymphatic distribution and in later stages of the disease pulmonary fibrosis.¹ The outcome of sarcoidosis in most patients is favorable except in those with extensive fibrosis. Occasionally, however, patients with sarcoidosis present with atypical findings on the chest imaging making the diagnosis more challenging. One pattern on the chest radiograph that is quite atypical for this disease has been called nodular sarcoidosis. Radiography and CT scanning show multiple nodules (less than 3 cm) or masses (more than 3 cm) that resemble metastatic cancer.

CASE REPORT

A twenty four year old previously healthy male presented to the hospital emergency room with four day history of constant, moderate intensity, right lower chest pain that was aggravated by movement and deep breathing. He reported similar pain on his left side two days later that radiated to his left arm. He denied palpitations, dyspnea, cough, fever, diaphoresis or recent weight loss. He denied smoking cigarettes.

On examination he was found to be afebrile and hemodynamically stable. There were no palpable lymph nodes. His chest was clear to auscultation and abdominal examination was benign. External genitalia were well developed with no masses. His neurological examination was within normal limits.

He had normal blood cell counts and indices as well as serum chemistries. An electrocardiogram was within normal limits and there was no elevation of cardiac enzymes.

A plain chest x-ray revealed multiple pulmonary nodules including a 2 cm opacity in the right lower lobe, a 1.5 cm opacity in the right upper lobe and a 3.2 cm opacity in the left upper lobe. (Figure 1) Computerized tomography verified these to be parenchymal nodules of soft tissue density without calcification. There was no associated cavitation, bony erosions or pleural effusions. Multiple enlarged lymph nodes were present including a 1.3 cm anterior para-tracheal node, and a two cm subcarinal node.

Further laboratory testing revealed a sedimentation rate of 10 mm/hr and C-reactive protein concentration of 3 mg/L. A screen for HIV was negative. Anti-nuclear antibody was positive with a dilution of 1:640 with nucleolar appearance. He was negative for RA, dS DNA, SSA, SSB and Scl-70 antibodies. Anti-proteinase-3 and antimyeloperoxidase antibodies were also negative. Angiotensin converting enzyme level was slightly elevated at 70 U/L (reference range nine to 67) as were SGOT at 41 IU/L (reference range 15-35) and SGPT at 58 IU/L (reference range 13-45). Alkaline phosphatase, bilirubin and INR were within normal limits. Mycoplasma IGM antibody titer was negative as was serum cryptococcal antigen. He had a negative PPD test and the sputum did not grow acid-fast bacilli (AFB), other bacteria or fungi.

A CT guided biopsy of the largest mass in the left upper lobe demonstrated multiple non-necrotizing granulomas with negative AFB, PAS

and GMS stains. Based on the clinical, radiological and histological features and exclusion of other suspected conditions, a final diagnosis of Stage II Sarcoidosis was made. Since the patient did not have any significant symptoms he was prescribed analgesics, provided information about his new diagnosis and scheduled for a follow up as an outpatient.

DISCUSSION

The vast majority of patients with sarcoidosis present with typical radio-



Figure 1. Chest X-ray revealed multiple opacities in the lungs including a 2 cm opacity in the right lower lobe abutting the pleura, a 1.5 cm focal density in the right upper lobe as well as a 3.2 cm rounded density in the left upper lobe.



Figure 2. Computed tomography of the chest showing the largest mass surrounded with smaller nodules (sarcoid galaxy sign).

graphic findings of bilateral hilar adenopathy, right paratracheal adenopathy and interstitial lung disease. Occasionally, patients will present with an atypical findings such as seen in our case. The differential diagnosis of large nodules and masses in the lungs is extensive and includes metastatic disease to the lungs, Wegener's granulomatosis, amyloidosis, fungal infections, lymphoproliferative diseases, lymphoma, pneumoconiosis and diffuse bronchoalveolar carcinoma.²

Most of the available knowledge concerning nodular sarcoidosis has been gathered from case reports and small case series.³⁻¹⁰ The incidence of this presentation is estimated to be approximately 1.5%. The majority of reported cases from the United States were in African-Americans, however, a review of 126 cases of pulmonary sarcoidosis in a Scandinavian cohort found a similar incidence.¹¹ In the largest series reported in the literature,¹² the majority of patients were females and smokers. Cough and shortness of breath were the most common presenting symptoms and chest pain was present in 51% of the patient's. Chest pain was the chief complaint and only complaint of our patient.

While nodular sarcoidosis is usually associated with multiple pulmonary nodules, occasionally a solitary mass may be the only finding.¹² Since most nodules or masses are sub-pleural in location, pleural irritation results in chest pain as seen in our patient. The masses may demonstrate air bronchograms and have been reported to cavitate. These masses are likely caused by the coalescence of smaller nodules within the interstitium. Nodular sarcoidosis however is a different entity than the conglomerate masses that can be seen in the upper lobes of some patients with ad-

vanced sarcoidosis resulting from intense peribronchovascular fibrosis. The latter is complicated by traction bronchiectasis and upper lobe volume loss.

Our patient was a young male with no significant past medical history. Testicular carcinoma was considered and the negative physical exam and pelvic CT scan were reassuring. Other malignancies were also considered as a transthoracic needle biopsy of the lung was promptly performed. It showed non-caseating granulomas consistent with sarcoidosis. Patients with nodular sarcoidosis in general have a favorable prognosis. Complete resolution of the masses or nodules can be expected either spontaneously or with oral corticosteroid therapy.

In summary, the presence of multiple masses and large nodules on a chest roentgenogram is often reason for concern. The differential diagnosis includes metastatic or primary malignancy. One benign disease that must be considered is nodular sarcoidosis. This diagnosis can be confirmed bronchoscopically with a transbronchial biopsy or by a CT guided transthoracic needle biopsy of the pulmonary lesion.

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