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Nodular Sarcoidosis Masquerading as Cancer

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ABSTRACT: Nodular lung disease is a rare pulmonary manifestation of sarcoidosis and resembles metastatic neoplasm disease. Nodular sarcoidosis is rare, varying from 1.6% to 4% of patients with sarcoidosis. Radiographic nodules measure from 1 to 5 cm in diameter that typically consist of coalescent granulomas. There is limited data on this form of sarcoidosis and its presentation can mimic primary or metastatic pulmonary neoplasms. Nodular sarcoidosis has a favorable prognosis, and resolution can be seen with oral corticosteroids. Herein, we present such a case of nodular pulmonary sarcoidosis with a lung nodule measured up to 6 cm.

KEYWORDS: Sarcoidosis, nodular sarcoidosis, noncaseating granuloma, hilar adenopathy, mediastinal adenopathy

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Introduction

Lung masses are usually suspected to represent a malignant process, especially if larger in size than 3 cm. Clinicians rely on radiologic characteristics of the mass to direct their diagnostic approach.[1–3] Positron emission tomography (PET) or integrated computed tomography and PET (PET/CT), when available, is often considered in the evaluation of lung nodules and masses. The positive predictive value of a positive PET scan exceeds 80%.[4] However, a positive PET scan can also be associated with inflammatory masses or pulmonary sarcoidosis.[5,6] The most common initial presentations of pulmonary sarcoidosis in adults are bilateral hilar adenopathy or reticulonodular opacities. Although the initial presentation of sarcoidosis as lung nodules is not uncommon, it rarely presents as large lung mass. We report a case of pulmonary sarcoidosis presenting with initial presentation as a lung mass.

Case Report

A 39-year-old African-American woman with a history of ulcerative colitis and tobacco abuse was referred for pulmonary consultation for atypical chest pain and abnormal chest x-ray (CXR). Her symptoms started 3 months prior to the presentation and her review of systems was unrevealing except for a decrease in appetite but without weight loss. Her physical examination was essentially normal and she had no lymphadenopathy or skin lesions. Initial CXR (Figure 1) found bilateral hilar adenopathy. Therefore, a chest CT scan was done which found a concerning mass. Therefore, a PET/CT scan of the chest was done and found mediastinal and hilar lymphadenopathy (Figure 2) and a right lower lobe lung mass (Figure 3) measured 4 cm × 3 cm mass all with significant fluorodeoxyglucose uptake.

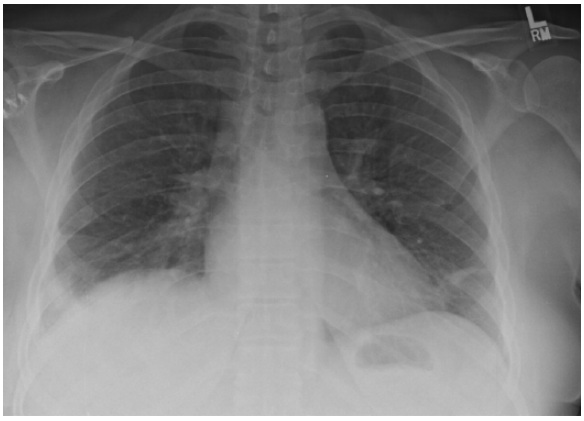

Figure 1. Portable chest x-ray. Bilateral hilar adenopathy.

Pulmonary function testing was performed and interpreted as follows: no defects, normal diffusion, and minimal air trapping. A CT-guided biopsy of the mass was consistent with nodular, noncaseous granulomas with background fibrous histiocytic infiltration and chronic inflammatory cells in the right lower lung. Based on these findings, the patient underwent video-assisted thoracoscopic surgery that revealed noncaseating granulomas consistent with sarcoidosis. Acid-fast bacilli stain, fungal, and tissue cultures were negative. A bronchoalveolar lavage was not performed. A diagnosis of sarcoidosis was made based on pathologic and radiologic evidence. The patient's symptoms resolved spontaneously without systemic glucocorticoids.

Discussion

Sarcoidosis is uncommon with an unknown etiology. In North America, the incidence of sarcoidosis is highest among black

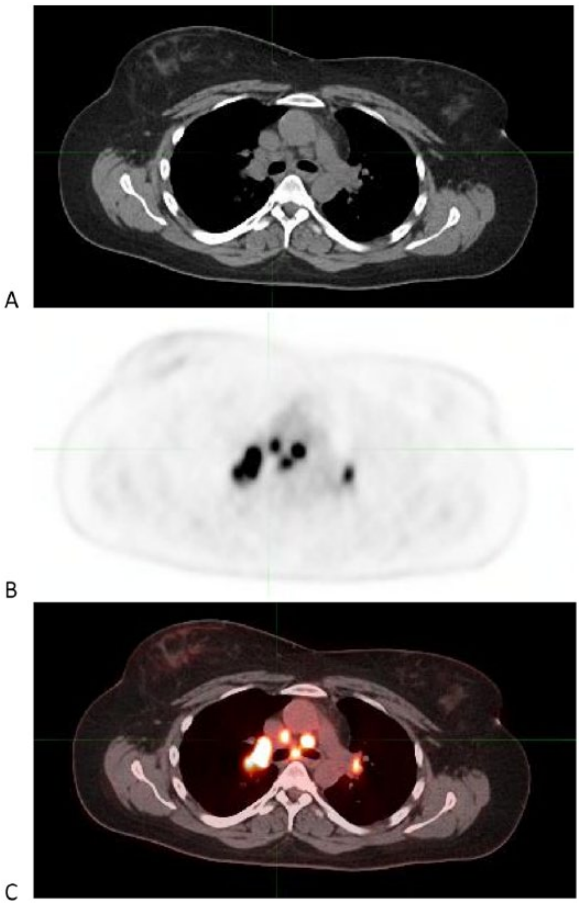

Figure 2. Positron emission tomography with computed tomographic (PET/CT) scan of mediastinal and hilar lung region. (A) Chest CT exhibiting mediastinal and hilar lymphadenopathy, (B) PET scan exhibiting uptake in mediastinal and hilar lymph nodes, and (C) fusion scan exhibiting uptake in mediastinal and hilar lymph nodes.

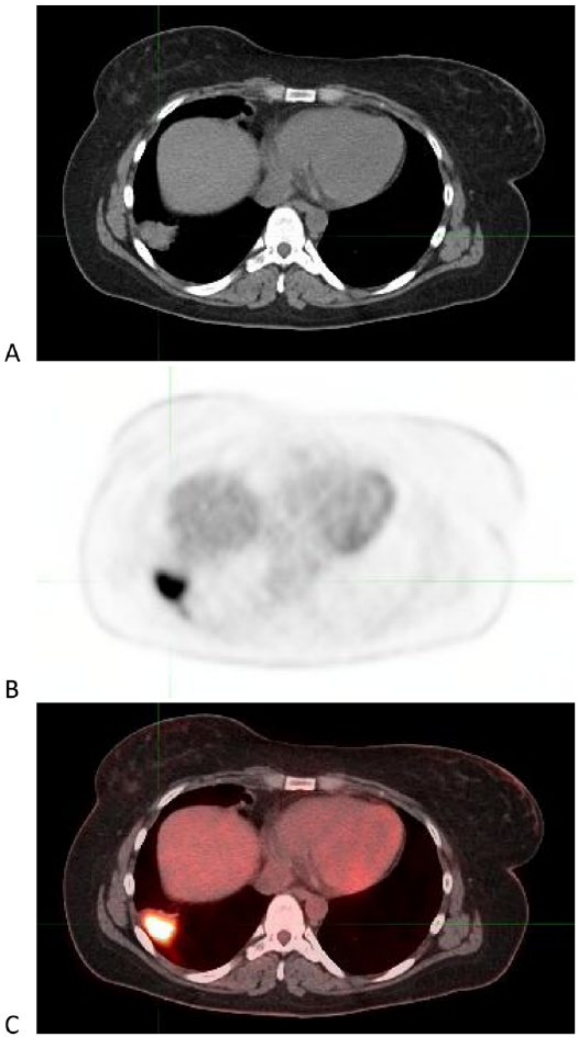

Figure 3. Positron emission tomography with computed tomographic (PET/CT) scan of lower lung fields. (A) Chest CT exhibiting a large right pleural density measured to be 4 cm × 3 cm, (B) PET scan exhibiting uptake in a large right pleural density, and (C) fusion scan exhibiting uptake in a large right pleural density.

women, whereas in Europe, it is highest among white women. Age of onset is between 30 and 45 years of age.[7] Typically, the diagnosis is straightforward based on pathognomonic signs and symptoms. But an atypical presentation often beguiles the medical community and can lead to misdiagnosis. Herein, we present an atypical case of sarcoidosis as a lung mass with no extrapulmonary manifestations.

Diagnosis of sarcoidosis requires 3 elements: (a) radiographic and clinical findings consistent with sarcoidosis, (b) exclusion of diseases that present similarly, and (c) biopsy of the most accessible lesion, demonstrating noncaseating granulomas on histopathology.[8] Sarcoidosis can present with atypical findings on chest radiography and CT scanning of nodules (<3 cm) or masses (>3 cm) that resemble primary or metastatic cancer.[9] Radiographic nodules measure from 1 to 5 cm in diameter that typically consist of coalescent granulomas.[10] These nodules usually tend to be peripheral. Imaging also can consist of bilateral hilar and mediastinal nodal enlargement, which is frequently seen in approximately 75% of the patients.[11] Despite these criteria, sarcoidosis is an everlasting diagnostic challenge.

Nodular sarcoidosis is an uncommon form of sarcoidosis and is prevalent in 2.4% to 4% of cases.[12] The first case of nodular sarcoidosis was reported by McCord and Hyman in 1952 with multiple bilateral nodules imitating metastatic disease.[13] Most of the patients with nodular sarcoidosis are African-American women between 20 and 40 years of age.[14]

The spontaneous resolution of the disease is common; however, up to 10% of the patients can progress to respiratory impairment and organ failure. There are no established criteria to determine the indications of treatment. The treatment is based on severity of symptoms, evidence of impairment in lung function, and disease with extensive fibrosis and bullae formation.[15] Oral glucocorticoids are the most commonly used medication in the treatment of sarcoidosis. Other alternative therapies such as immunosuppressive and cytotoxic agents have been suggested, but there is limited evidence regarding

efficacy of these agents. Nodular sarcoidosis presenting as lung mass is exceedingly rare. Most of the identified patients are women and smokers. These solitary masses are believed to be a coalescence of smaller nodules and prove to be even more misleading and suggestive of a primary cancer. Patients with nodular sarcoidosis have a favorable prognosis with significant improvement in radiographic findings with systemic corticosteroids.[16]

Author Contributions

AJS, NKS, AS, and MT contributed substantially to the authorship of this case report. AS and MT were directly involved in the patient's care.

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