Multiple Lung Masses, Pneumothorax, and Psychiatric Symptoms in a 29-Year-Old African-American Woman*

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MRI=magnetic resonance imaging

A 29-year-old African-American woman was admitted to the hospital with a 1-day history of mild pleuritic chest pain. The patient had a 3-month history of nonproductive cough, progressive dyspnea, and low-grade fever, accompanied by a 6.75-kg weight loss. She had exhibited a flattened affect and slow mentation for the past several months and had been prescribed haloperidol by a psychiatrist for “undifferentiated schizophrenia.” A native of Los Angeles, the patient had smoked one pack of cigarettes per day for the past 15 years. She did not abuse drugs or alcohol and had not traveled outside of the Los Angeles area.

Physical Examination


Laboratory Findings


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Hospital Course

Fiberoptic bronchoscopy with transbronchial biopsy was performed that established the diagnosis. The patient was discharged from the hospital without specific treatment owing, in part, to her psychiatric history. One month later, the patient was readmitted to the hospital with a 40% pneumothorax of the right lung (Fig 3).

What diagnosis most likely can explain the patient’s clinical presentation?
Diagnosis: Nodular sarcoidosis with pneumothorax and probable neurologic involvement

Nodular sarcoidosis is noted in about 2 to 4% of all cases of pulmonary sarcoidosis. In the United States, the predominance of cases involve young African-American adults. The course, usually benign with the disease limited to the chest, has a tendency toward regression with corticosteroid therapy.

When a young individual presents with bilateral lung masses and adenopathy, several diagnostic possibilities should be considered. The important feature of this entity is its resemblance to malignant chest lesions such as metastatic cancer and lymphoma. Of the malignant lesions, breast cancer, germ cell tumor, hypernephroma, sarcoma, and lymphoma are the most likely to consider with the radiographic presentation. Infectious granulomatous disease, such as tuberculosis or coccidioidomycosis, and noninfectious entities such as Wegener’s granulomatosis also are diagnostic possibilities. The presence of extensive mediastinal adenopathy without hoarseness or superior vena cava syndrome tends to support a benign etiology.

The finding of an air bronchogram within lung masses provides an important diagnostic clue, as it strongly suggests lymphoma or benign aggregates of lymphoid cells, as is found in sarcoidosis. The diagnosis of nodular sarcoidosis can be established either by transbronchial biopsy or by percutaneous fine-needle aspiration if the lesions are peripheral. Pneumothorax is rare in sarcoidosis and is probably due to rupture of a subpleural bleb or necrosis of a sarcoid granuloma. After reexpansion of the lung, recurrence of pneumothorax can, in most cases, be prevented by corticosteroid therapy.

Neurologic involvement is generally found in about 5% of all cases of sarcoidosis, but the incidence has varied from 2 to 16% in different series. About half of these patients have neurologic symptoms as their presenting complaint. Most, however, will either have or later develop evidence of disease elsewhere. Sarcoid granulomas can affect virtually any part of the nervous system, producing protean neurologic manifestations. These include cranial neuropathy, meningoencephalitis, space-occupying lesions, hypothalamic and pituitary abnormalities, myelopathy, and peripheral neuropathy.

Cranial neuropathy is the most prevalent lesion in neurosarcoidosis. Although facial palsy predominates, any of the cranial nerves can be affected, either alone or in combination, thus producing a variety of symptoms. Visual deficit, deafness, dysphagia, and hoarseness have all been reported. Peripheral neuropathy can present as mononeuritis, mononeuritis multiplex, and various sensory or motor polyneuropathies. Isolated cranial or peripheral neuropathy tends to have a favorable response to steroids and a relatively good prognosis. Involvement of the hypothalamic and pituitary region is also well recognized, producing diabetes insipidus and occasionally other endocrinopathies. Evidence of meningeal involvement is common on MRI, by cerebrospinal fluid study, and at autopsy, but...
Clinical meningitis is infrequent. Involvement of the ependymal lining may produce hydrocephalus. Sarcoid granulomas may enlarge to mimic central nervous system tumors. Involvement of the cerebellum and the spinal cord have rarely been reported.

Encephalopathy, ranging from mood disturbance (apathy, anxiety), personality change, and deteriorating intellect, to dementia and psychosis, has been recognized with increased frequency. Symptoms of mental disorders have usually been in association with meningencephalitis or space-occupying lesions. In a few cases, results of imaging and cerebrospinal fluid studies have been normal, possibly owing to the small size of the sarcoid granulomas or the lack of sensitivity in detecting lesions in this particular anatomic location.

Computed tomography and MRI have proved invaluable in the evaluation of neurosarcoidosis. A head CT with contrast can show intracranial lesions, hydrocephalus, and even basal meningeal involvement, but sensitivity suffers in the cerebellum, brain stem, hypothalamus, and pituitary. Magnetic resonance imaging enhanced with gadolinium appears to have a sensitivity exceeding 90% and is the imaging modality of choice in neurosarcoidosis. Neither CT nor MRI findings are specific for neurosarcoidosis.

Corticosteroid therapy remains the cornerstone of treatment of neurosarcoidosis. In contrast to pulmonary sarcoidosis, the initial dose tends to be higher, the treatment duration longer, and the response generally poorer. While corticosteroid therapy can improve psychiatric symptoms due to neurosarcoidosis, it is also known to precipitate or exacerbate neuropsychiatric syndromes, and, thus, presented a dilemma in managing the present patient with known psychosis and a normal MRI. When the present patient was admitted to the hospital with a pneumothorax, she had thoracic aspiration with prompt resolution; corticosteroid therapy was then begun at an initial dose of prednisone, 20 mg/d, and the drug treatment was tapered to 5 mg/d over 3 months. A chest radiograph showed marked improvement (Fig 4). There was no recurrence of pneumothorax at 6-month follow-up. Haloperidol therapy was discontinued, and her psychiatric symptoms were substantially improved with prednisone therapy, suggesting a diagnosis of neurosarcoidosis.

**Clinical Pearls**

1. Nodular sarcoidosis, observed in less than 5% of all cases of pulmonary sarcoidosis, usually affects young African-American adults.
2. Nodular sarcoidosis tends to be limited to the thorax and to respond to corticosteroid treatment.
3. The finding of an air bronchogram within lung masses on CT scan suggests lymphoma or sarcoidosis and argues against carcinoma.
4. Pneumothorax is rare in sarcoidosis; recurrence can usually be prevented with corticosteroid therapy.
5. Clinically recognizable neurologic involvement occurs in approximately 5% of patients with sarcoidosis, with neurologic symptoms as the initial complaint in nearly half of the patients.

**Suggested Readings**


Mathuswamy P, Chakravarty A, Lopez-Majano V. Nodular sarcoidosis is a 'limited' form of sarcoidosis. Sarcoidosis 1994; 11(suppl):129-86


Scott TF. Neurosarcoidosis: progress and clinical aspects. Neurology 1993; 43:8-12