Unilateral Pulmonary Hilar Tumor Mass: Is It Always Lung Cancer?

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Abstract

Sarcoidosis is a multisystem inflammatory disease of unknown etiology, characterized by noncaseating epithelioid cell granulomas. In sarcoidosis, the most common radiological findings are mediastinal and bilateral hilar lymph node enlargement. We present a case of sarcoidosis with a rare radiological aspect of pulmonary hilar tumor mass.

A 54-year-old female patient, active smoker (40 packs/year), with a history of cutaneous lupus, was admitted in our institute for progressive dyspnea and dry cough. At admission physical examination and laboratory tests were normal. Pulmonary function tests diagnosed an obstructive syndrome. Chest X-ray showed a tumor mass of the right pulmonary hilum. Transbronchial biopsy was nondiagnostic. HRCT-scan showed a tumor mass in the right hilum, which raised the suspicion of a lung cancer. PET-CT scan revealed a high metabolic activity of the tumor mass and of a paratracheal right lymphadenopathy. Lymph node biopsy by mediastinoscopy showed noncaseating epithelioid-cell granulomas, sustaining the diagnosis of sarcoidosis. The outcome was favorable, with spontaneous remission without treatment, but with a relapse that responded after systemic corticotherapy.

In conclusion, even if a tumor mass in the pulmonary hilum is highly suggestive of lung cancer, a positive diagnosis should be made only after histological examination, because other benign conditions, like sarcoidosis, could have such an aspect.

Keywords: sarcoidosis, lung cancer, tumor mass

Case Report

Sarcoidosis is a multisystem inflammatory disease of unknown etiology, characterized by noncaseating epithelioid cell granulomas (1,2). Any organ may be involved, but more than 90% of patients have thoracic manifestations (1,3). Although radiological findings may vary (4), bilateral hilar lymph node enlargement seems to be the most common aspect (between 50% and 85% of patients) (5). We present a case of sarcoidosis with a rare radiological aspect consisting of unilateral hilar tumor mass which made difficult the differential diagnosis with the lung cancer.

A 54-year-old Caucasian female patient, with a 40 packs/year smoking history, was admitted in our hospital in November 2007 for a history of progressive dyspnea and dry cough,
which gradually worsened one month prior to admission. Patient had no previous medical history, except a possible cutaneous lupus at age 34 which resolved without treatment. Physical examination was normal (none of the following: fever, peripheral lymphadenopathy, arthritis, peripheral edema, pulmonary rales; the blood pressure was 140/80 mmHg, the pulse was 78 beats/minute, the respiratory rate was 16 breaths/minute, and the oxygen saturation was 97% while breathing ambient air). Pulmonary function tests identified an irreversible obstructive syndrome which conducted to a COPD diagnosis. Post-bronchodilator spirometry values were: FEV1/FVC ratio 0.68, FVC 1.88L (68.2% predicted), FEV1 1.28L (55% predicted). The chest X-ray showed a tumor mass in the right pulmonary hilum (Figure 1). Results of laboratory tests, including the serum levels of angiotensin-converting enzyme, α1-antitripsine, rheumatoid factor, antinuclear antibodies, complement, tumor markers (neuron-specific enolase, carcinoembryonic antigen, CA-125), were normal. No abnormalities were found on cardiac ultrasonography. Bronchoscopy showed only a flattening of the emergency of the right upper lobe bronchus, but no bronchial tumor. Bronchoalveolar lavage showed an elevated total cell number (18x10^6 cells), with the predominance of lymphocytes (37%) and neutrophils (5.4%). Smears for acid-fast bacilli were negative, all bacteriological cultures were negative, and no tumor cells were found. Transbronchial biopsy of the lymphadenopathies from the right upper lobe revealed only mild fibrosis, without granulomatous lesions or cancer cells). Thoracic HRCT-scan (Figure 2) showed a tumor mass in the right pulmonary hilum with an aspect highly suggestive of lung cancer, lymphadenopathy of 3.2 cm in Barety’s space, small lymphadenopathies in prevascular space, one dense nodule 1.8 cm in diameter in the left suprarenal gland, without lung parenchymal lesions or abdominal lymphadenopathies. At this point, the lung cancer seemed the most likely diagnosis and a mediastinoscopy was recommended to the patient, but the patient refused it. In April 2008 she returns to our hospital. During this period she underwent different investigations: a PET-CT scan with a high metabolic activity on the right pulmonary hilum and in the right paratracheal lymphadenopathy, but no high metabolic activity of the suprarenal gland.
nodule (Figure 3), and a biopsy of a supraclavicular lymphadenopathy with noncaseating granulomatous tissue raising the suspicion of tuberculosis (no cultures from the biopsy were performed). Bronchoscopy and bronchoalveolar lavage were repeated and had the same aspect. At that time, the patient accepted mediastinoscopy with mediastinal lymph node biopsy which revealed noncaseating epithelioid-cell granulomas (Figure 4), confirming the diagnosis of sarcoidosis. A follow-up evaluation 3 months later revealed a favorable outcome with spontaneous regression of the unilateral hilar tumor mass (Figure 5). In October 2009, the patient developed a relapse of sarcoidosis with severe dyspnea and a HRCT-scan showing the right hilar tumor with partial atelectasis of the right upper lobe (Figure 6), with a good clinical and radiological outcome after a 3 months course of systemic corticotherapy (Figure 7).

**DISCUSSION**

Sarcoidosis, an immune-mediated granulomatous disease, remains a diagnosis of exclusion (6). Typical imagistic features (i.e. bilateral hilar lymphadenopaties) are frequently attributed to sarcoidosis, but the disease has a widerange of radiological aspects. Although the lungs, the mediastinal and bilateral hilar lymph nodes are the most common sites of involvement (6,7), sarcoidosis wasn’t take into account in our case from the beginning, due to the atypical clinical and radiological findings. Having a 40% to 90% range of diagnosis, the flexible fiberoptic bronchoscopy with transbronchial lung biopsy is the initial recommended procedure in patients suspected of sarcoidosis (8) but, unfortunately, in our case it was nondiagnostic. The PET-CT scan revealed a high metabolic activity in the hilar tumor mass, which can appear in lung cancer and in several other conditions with active granuloma formation, such as: sarcoidosis, tuberculosis, nontuberculous mycobacterium granuloma, and fungal infections. Only after the patient’s agreement for mediastinoscopy we could analyse the histology of the mediastinal lymph nodes, making the differential diagnosis between sarcoidosis and lung cancer. Regarding the outcome, our patient was part of the 60-70% of people diagnosed with sarcoidosis who have a spontaneous remission of the disease (3).
The presented case has some particularities. First, we cannot say for sure if the functional obstructive syndrome is due to COPD in a smoker patient or to sarcoidosis, in which 30% of patients develop an obstructive syndrome (9) (probably both pathologies had contributed to it). Second, since cutaneous lesions are common in sarcoidosis, but nonspecific (10) the cutaneous lupus diagnosed at age 34 could have been the first manifestation of sarcoidosis in our patient. But the most important aspect was the radiological aspect with a right hilar lung tumor, this pattern being described only in 1 to 3% of patients with sarcoidosis (5).

In conclusion, even if a tumor mass in the pulmonary hilum is highly suggestive of lung cancer, a positive diagnosis should be made only after histological examination, because other benign conditions, like sarcoidosis, could have such an aspect.

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REFERENCES


Abbreviations list
COPD = chronic obstructive pulmonary disease
FEV1 = forced expiratory volume in the first second
FVC = forced vital capacity
HRCT-scan = high-resolution computed tomography
PET-CT scan = positron emission computed tomography

FIGURE 7. Chest radiography showing regression after corticotherapy for relapse.