INFLAMMATORY PSEUDOTUMOR OF THE LUNG IN A COAL MINER WITH PNEUMOCONIOSIS

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Abstract: Inflammatory pseudotumors of the lung are uncommon and etiologically diverse lesions that often present as solitary masses in the lung. It may be difficult to distinguish these lesions from more commonly encountered lung neoplasms. Inflammatory pseudotumors can also occur in other organs, but the lung is most commonly involved. We describe a 63-year-old male coal miner with a 40-year history of dust exposure, who had a large right middle lobe mass on chest roentgenograms, with slow growth over the 7 years prior to admission. Repeated transthoracic echo-guided biopsies of the mass were indicative of an inflammatory and reactive process. The radiographic, histologic, and clinical findings indicated a diagnosis of inflammatory pseudotumor. The patient refused surgical intervention and was regularly followed at our outpatient clinic. Follow-up chest roentgenograms for 1 year revealed that the tumor size was stable. This case suggested that inflammatory pseudotumor, although uncommon, should be included in the differential diagnosis in a patient with pneumoconiosis and a solitary mass in the lung.

Inflammatory pseudotumors of the lung are uncommon. Since the first description of an inflammatory pseudotumor of the lung by Brunn in 1939 [1], the lesions have been variously referred to as plasma cell granuloma [2], inflammatory myofibroblastic tumor [3, 4], and inflammatory myofibrohistiocytic proliferation [5]. The lesions are composed of chronic inflammatory cells, such as lymphocytes and plasma cells, and fibroblasts [2]. Although similar lesions have been documented in other organs, the lung is by far the most commonly involved organ. However, these lesions are uncommon even in lungs.

The inflammatory pseudotumor often presents as a solitary mass in the lung. Therefore, it may be difficult to differentiate from more commonly encountered lung neoplasms on imaging. The diagnosis thus depends on microscopic examination of tissues.

Here, we describe a case of inflammatory pseudotumor of the lung in a patient with pneumoconiosis.

Case Report

A 63-year-old man visited our clinic due to cough and dyspnea of 2 years’ duration. He had worked as a coal miner for 40 years until his retirement 9 years ago. He was a non-smoker. Fever, chills, recent body weight loss, and trauma were denied. Physical examination revealed normal and stable vital signs. Breathing sounds were clear without wheezing. Other physical examinations revealed essentially normal findings. The results of laboratory tests, including complete blood count and routine blood chemistry, were also within normal limits. The results of sputum smears for acid-fast bacilli were repeatedly negative, as was culture for mycobacteria. An 8.5 x 11.6-cm right middle lobe mass, which was homogeneous in density and had well-defined margins, was observed on chest roentgenograms. The remainder of the lung showed a diffuse reticulonodular pattern and progressive massive fibrosis (Fig. 1B). Sequential roentgenograms revealed slow growth of the mass over a 7-year period (Fig. 1).
Inflammatory Pseudotumor of the Lung

Fig. 1. A) Chest roentgenogram taken on February 20, 1997, showing a large lesion over the right lung. B) Larger lesion over the same location on June 30, 1998.

Computerized tomography of the thorax also confirmed the presence of a right middle lobe mass associated with bilateral homogeneous suprahilar soft tissue density (Fig. 2), which was interpreted as progressive massive fibrosis. Pulmonary function test showed a forced vital capacity (FVC) of 2,500 mL (90% predicted value); a forced expiratory volume in 1 second (FEV₁) of 1,540 mL (72% predicted value); an FEV₁/FVC ratio of 62%; a total lung capacity of 3,740 mL; and a residual volume of 1,240 mL.

Bronchoscopy revealed patchy anthracotic pigmentation of the airway mucosa and more than 90% narrowing of the right middle lobe bronchus by external compression.

Transbronchial lung biopsy of the right middle lobe was performed and histology showed anthracosilicosis without evidence of neoplasm. Six days later, transbronchic echo-guided lung biopsy of the tumor was performed. Histologic analysis showed haphazardly arranged, dense hyalinized collagenous bands of varying thickness, as well as fibroblasts and scattered lymphocytic infiltrates, indicative of an organizing inflammatory lesion (Fig. 3). Aspiration cytology examination of a dried smear revealed reactive fibroblast-like cells (Fig. 3A). These cells differed from macrophages in that they had less cytoplasm and slightly irregular cytoplasmic profiles. The presence of round vacuoles of varying sizes throughout the smear including extra-
cellular areas indicated that the vacuoles were probably artifacts caused by unclean oily glass. Phagocytized particulate granules, dust or carbon, were not seen within the cytoplasm. The nuclei were similar to those of young fibroblasts observed in the biopsies (Fig. 3D). Repeat transthoracic echo-guided lung biopsy yielded the same results and pneumoconiosis with inflammatory pseudotumor was diagnosed. The patient refused surgical intervention and was regularly followed at our outpatient clinic. Follow-up chest roentgenograms for 1 year revealed that the tumor size was stable.

Discussion

Inflammatory pseudotumors are uncommon inflammatory lesions that have been observed in various organs, including the lung [6], liver [7], kidney, urinary tract, and gastrointestinal tract [8]. These lesions, which are inflammatory and non-neoplastic in nature, may mimic neoplasms in clinical presentation [9]. Histologically, the lesions are comprised of fibroblasts, inflammatory cells, and varying quantities of collagenous tissues. Some of the fibroblasts may contain abundant intracytoplasmic actin filaments, and are often referred to as myofibroblasts [10, 11].

Because of the inflammatory nature of the lesions, they may occasionally regress spontaneously [12]. Recurrence following surgical resection has been recorded [13]. Gram-negative bacteria, especially Escherichia coli, have often been associated with pseudotumors involving the gastrointestinal and urinary tracts [14]. In the respiratory system, inflammatory pseudotumors have been documented in immunodeficiency [15], trauma patients, and patients with aspiration pneumonia and miscellaneous infections such as Q fever [2, 16].

Recent studies have suggested that inflammatory pseudotumor may involve monoclonal or polyclonal
proliferation of dendritic cells of the lymphoid tissue [4, 17]. Rare cases of monoclonal proliferation [17] and sarcomatous transformation [4, 18] of pseudotumors have been described. Malignant fibrohistiocytoma may be difficult to distinguish from inflammatory pseudotumor during certain stages of evolution [4, 19]. Therefore, rare malignant behavior of seemingly benign inflammatory pseudotumors may be encountered [4, 19].

Radiographically, no specific findings are helpful in the diagnosis of the disease [20, 21]. Agrons et al reported that, on chest roentgenograms, 87% of patients had solitary peripheral lesions (masses or nodules), of which 60% occurred in the lower lobe [22]. On chest CT scan, most lesions were sharply circumscribed; in only one of 17 patients were the borders ill defined. Calcification was uncommon. Magnetic resonance imaging showed heterogeneous lesions with signal intensity slightly greater than that of skeletal muscle on T1-weighted images. High signal intensity was noted on T2-weighted images.

Golbert and Pletnev reported that the incidence of inflammatory pseudotumors of the lung was 0.7% in 1,075 lung tumors [23]. These tumors occur in individuals of all ages but are considered to be the most common tumor-like lung lesions in children younger than 16 years [2]. In adults, they tend to occur in patients of 70 years or older [6].

Clinical presentation of the pulmonary inflammatory pseudotumors is nonspecific. Berardi et al reported that only 26.2% of patients were symptomatic [24]. The most common symptoms included productive cough, fever, pain, and rarely, hemoptysis. The results of laboratory examinations were nonspecific [6].

Complete excision of the mass with preservation of the normal lung tissue is a reliable method for diagnosis and treatment [6]. Surgical excision is the mainstay of treatment. Usually, wedge resection is adequate if removal is complete. However, lobectomy or even pneumonectomy is occasionally necessary for complete resection. Repeat resection should be performed if pseudotumors recur [25].

Although a definite cytologic diagnosis of inflammatory pseudotumor can only be made by histologic examination and not by fine-needle aspiration cytology, the cytologic features can be distinguished from other, more usual, benign pulmonary lesions or malignancy. Transthoracic fine-needle aspiration cytology of pulmonary inflammatory pseudotumors shows moderately to poorly cellular smears containing a mixture of chronic inflammatory cells, epithelial cells, and tissue fragments, including fibroblasts and macrophages [26]. The cytologic atypia are minimal.

Pathologic findings of inflammatory pseudotumor of the lung may show a spectrum of histologic features [27]. They include proliferation of spindle-shaped fibroblasts and infiltration of lymphocytes, histiocytes, lymphoid hyperplasia with lymph follicles, and intraalveolar fibrosis at the edge of the tumor. On the basis of the predominant histopathologic features, three major patterns are identified [27]: organizing pneumonia, fibrous histiocytic pattern, and lymphoplasmocytic pattern. The organizing pneumonia pattern contains airways filled with plump fibroblasts and foamy histiocytes. Fibroblasts occasionally grow into fibrin and inflammatory cells in alveoli, alveolar ducts, and bronchioles. The fibrous histiocytic pattern is characterized by spindle-shaped myofibroblasts arranged in whorls. The lymphoplasmocytic pattern contains a mixture of lymphocytes, plasma cells, and histiocytes with only minimal fibrous connective tissue.

In our patient, echo-guided biopsies were used to establish the diagnosis. The potential for insufficient tissue sampling cannot be completely excluded. Therefore, surgical excision was advised but refused by the patient.

A few other non-neoplastic, reactive processes were also considered in the differential diagnosis. At the top of the list was coalworker’s pneumoconiosis with progressive massive fibrosis (PMF). In PMF, the mass lesion usually presents with a well-defined lateral border that parallels the thoracic cage, with a zone of emphysematous lung between it and the chest wall [28, 29]. It is usually found in the upper lung field. Foci of degenerating or necrotic collagenous tissues are often seen in biopsies or aspiration samples [29]. Such histologic or cytologic features were absent in the current case. Caplan’s syndrome consists of the presence of nodules associated with rheumatoid arthritis superimposed on a background of inorganic dust exposure [29], making it another disease worthy of considering. Arthritis or rheumatoid nodules were absent in our patient and features of rheumatoid nodules were not observed in biopsies or aspiration cytology. The necrotic center of rheumatoid nodules can be aspirated with relative ease, but this was not observed in our patient. The clinical, radiographic, and morphologic features of our patient favored the diagnosis of an inflammatory pseudotumor. Although coal dust exposure may have played a role in the formation of the pseudotumor in our patient, the exact etiology remains unclear.

References