CASE REPORT

Inflammatory Myofibroblastic Tumor of the Lung

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ABSTRACT

Inflammatory myofibroblastic tumor of the lung is a rare condition, with a reported incidence between 0.04 - 1.2% of all tumors of the lung. We present a case of inflammatory myofibroblastic tumor of the lung. A 61-year man presented to the outpatient department complaining of cough and blood-streaked sputum for 5 days. The computed tomography scan of the chest demonstrated a 4.5 x 4 cm, calcified pulmonary mass in the anterior segment of the right upper lobe. Bronchoscophy and computed tomography-guided transthoracic fine needle aspiration was inconclusive. The tumor was removed via wedge resection. Histological and immunohistochemical findings were consistent with inflammatory myofibroblastic tumor of the lung.

Key Words: *Lung. Inflammatory myofibroblastic tumor. Surgery.*

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare disease, accounting for 0.4 - 1.2% of all lung tumors.[1] This tumor is composed of a myofibroblastic spindle cell population accompanied by an infiltrate of variable numbers of inflammatory cells including lymphocytes, plasma cells, eosinophils, immunoblasts, and fibrous tissue.[2] Its etiopathogenesis is unknown, but recently, several reports have shown that IMT is true neoplasm rather than a reaction process.[2,3] IMTs of the lung mimic malignant process clinically and radiographically.[4] They are most commonly parenchymal in location.[2] Endobronchial IMT is rare, with a prevalence between 0 and 12%.[2,3] Because preoperative diagnostic methods are usually inconclusive for diagnosis, surgery is often required for both diagnosis and treatment.[1,5] The prognosis of patients who undergo surgical radical resection is usually excellent.[1]

We report a 61-year male patient with IMT of the lung, diagnosed and treated with wedge resection.

CASE REPORT

A 61-year man presented to the outpatient department complaining of cough and blood-streaked sputum for 5 days. He was smoker. There was no previous history of any respiratory or other diseases. A physical examination revealed no abnormalities. Chest X-ray showed a 4 cm mass shadow in the right perihilar region (Figure 1). The findings from physical examination were unremarkable. Erythrocyte sedimentation rate was 100 mm/first hour. Serum C-reactive protein level was 162 mgr/L. Other routine laboratory values were within normal ranges. The computed tomography scan of the chest demonstrated a well-demarcated, solitary, non-homogeneous, 4.5 x 4 cm, calcified mass in the anterior segment of the right upper lobe (Figure 2). There were multiple lymphadenopathies in the mediastinum. Flexible bronchoscopy revealed normal endobronchial appearance. Computed tomography-guided transthoracic fine needle aspiration was inconclusive. The maximum standardized uptake values (SUVmax) of the pulmonary mass and mediastinal lymphadenopathies on FDG PET-CT were 13.2 and 8.9, respectively. Mediastinoscopy revealed a diagnosis of reactive hyperplasia and anthracofibrosis of lymph nodes.

A standard right postero-lateral thoracotomy was performed. The mass was palpable in the anterior segment of the right upper lobe. Frozen section analysis indicated a benign lesion with clear margins. The tumor was removed via wedge resection. Grossly, the resected tumor was a single round, well circumscribed, grey-white in colour mass with calcification, measuring 4.5 cm x 4 cm in size. Microscopic examination revealed a proliferation of spindle cells showing myofibroblastic differentiation arrayed in fascicles, admixed with an inflammatory infiltration containing lymphocytes, eosinophils, and plasma cells (Figure 3). Immunohistochemical analysis showed positive staining for vimentin and smooth muscle actin, and negative staining for desmin, S-100, and CD-34. These findings were consistent with inflammatory myofibroblastic tumor of the lung. The patient has survived without recurrence for 18 months to date after the operation.

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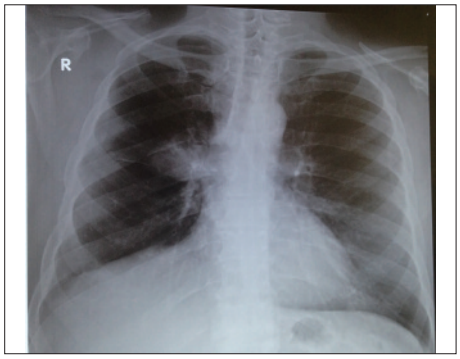

Figure 1: Chest X-ray shows a 4 cm mass shadow in the right perihilar region.

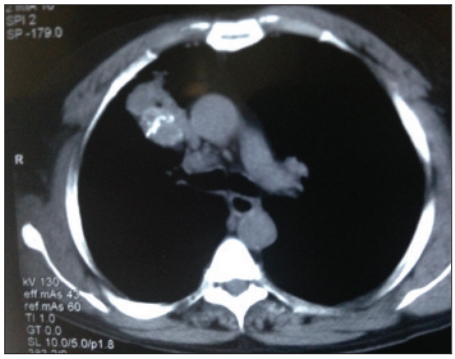

Figure 2: The computed tomography scan of the chest demonstrated a 4.5 x 4 cm, calcified pulmonary mass in the anterior segment of the right upper lobe.

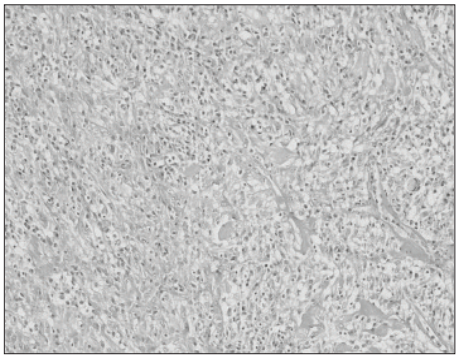

Figure 3: Microscopic examination shows a proliferation of spindle cells showing myofibroblastic differentiation arrayed in fascicles, admixed with an inflammatory infiltration containing lymphocytes, eosinophils, and plasma cells (Hemotoxylen and Eosin x 10)

DISCUSSION

Inflammatory myoblastic tumor is a rare mesenchymal lesion.[6] Although it can occur at a wide variety of other sites including abdomen (mesentery, omentum, liver, spleen, kidney, pancreas, stomach, ileum, rectum), retroperitoneum, pelvis (bladder, uterus), upper respiratory tract, pharynx, mediastinum, esophagus, orbits, intracranial space, heart, lymph nodes, salivary glands, epididymis, extremities, and muscles.[3-6] Pulmonary location is the most common site.[7,8] This tumor constitutes 0.04 - 1.2% of all tumors of the lung.[1] IMT is the most common isolated primary neoplasm in the lung in children younger than 16 years.[6] The nature of this tumor is controversial. According to some authors, IMT represents an immunologic/inflammatory response to an infectious or non-infectious intervention or insult. According to others, it is a true neoplasm. This theory is supported by detection of cases of IMT with local invasion, distant metastases or multi-centric disease. Additionally, recent cytogenetic studies have supported that IMT is a true neoplasm.[1-5]

IMTs can occur at any age, however, approximately 60% of these tumors occur in the patients younger than 30 years of age. They do not show gender predilection.[2] Patients generally have non-specific symptoms. Common symptoms include cough, dyspnea, hemoptysis, fever, pleuritic pain, and respiratory infections.[1,7] However, patients are asymptomatic in 30 to 70 % of cases, and the tumor is incidentally detected on chest radiological examinations performed for other reasons.[1,5] Clinical presentation of IMT may depend on location. Parenchymal IMTs are usually asymptomatic, but endobronchial and larger/invasive parenchymal tumors may cause pulmonary symptoms.[2-4] A history of upper respiratory tract infection or pneumonia is reported in approximately 30% of cases.[7] In this case, the tumor was located in lung parenchyma. Cough and hemoptysis were the main symptoms, and the patient had no history of previous respiratory infection.

Radiological features are variable and non-specific. Computed tomographic scan shows a solitary nodule or mass in 90% of patients.[7] The lesion can be homogeneous or heterogeneous with or without clear margins.[5,6] Calcification or cavitation is very infrequent.[7] FDG-PET scan demonstrates an uptake, similar to that of malignant tumors.[9] In this patient, computed tomography scan showed a well-demarcated, solitary, non-homogeneous, and calcified mass and the tumor had a high FDG uptake. Since clinical and radiological manifestations of IMT of the lung are diverse and non-specific, the diagnosis of this tumor requires histological and immunohistochemical examinations to exclude alternative diagnosis.[5,7] Pathological differential diagnosis of IMT is multifarious because of its variable cellular admixture. It includes lymphoid hyperplasia, malignant lymphoma, pseudolymphoma, plasmacytoma, malignant fibrous hystiocytoma, sarcomatoid carcinoma, sclerosing hemangioma, sarcoma, primary lung cancer, fibrosis, organized pneumonia, angiomyofibroblastoma, and desmoid fibromatosis.[5,7,10] Pre-operative diagnosis of this tumor is difficult. Bronchoscopy and transthoracic fine-needle aspiration are often inconclusive. In most cases, precise diagnosis is usually established by surgical resection.[1,5,10] The diagnosis of IMT was established by surgical resection in this patient.

The treatment of choice for IMT is surgical resection. Whenever possible, wedge resection should be considered as the first-line treatment. If needed, lobectomy or pneumonectomy can be performed to ensure radical resection. Enbloc resection may be needed in cases of chest wall invasion or carina or main bronchus, pericardium or diaphragm involvement.[1,6-8] Complete surgical resection is advocated to prevent recurrence.[1] Local recurrence rate of IMT of the lung after resection have been reported to be between 6.6% and 13%. It occurs usually in the setting of incomplete resection.[6] The effectiveness of steroids, chemotherapy or radiation is uncertain. Corticosteroid therapy has

been proposed in the case of inoperable patients, for concurrent cardio-respiratory disease, for functionally inoperable patients, for unresectable lesions or in case of recurrence.[1,8] Chemotherapy or radiation therapy has been reserved for patients in whom resection is prohibitively morbid or technically unfeasible, or in patients who had incomplete resections, local recurrence or metastatic disease.[1,6,8] The prognosis of patients who undergo radical resection is excellent. Survival rates at 5 and 10 years were reported to be 91% and 77%, respectively.[1] Nevertheless, relapse and disease-related deaths can occur even many years after.[7] In this case, the tumor was removed via wedge resection. The patient has survived without recurrence for 18 months to date after the operation.

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