Primary endobronchial mucosa-associated lymphoid tissue lymphoma presenting with hemoptysis: A case report

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Abstract

Primary pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma is an extremely rare disease, which can involve the lung parenchyma or bronchi. The most common findings of primary pulmonary MALT lymphoma are a solitary nodule or mass with or without air bronchograms by chest roentgenogram or computerized tomography of the chest. We describe a case of primary pulmonary MALT lymphoma presenting with normal radiological appearance and intermittent hemoptysis. Bronchoscopic examination showed a plaque lesion located on the lower trachea just above the carina.
Introduction

Primary pulmonary MALT lymphoma is a rare extranodal lymphoma that is usually of the low-grade B-cell type and is considered to arise from mucosa-associated lymphoid tissue (MALT) of the bronchus, which is histologically distinct from true intrapulmonary lymph nodes. MALT-associated malignant lymphomas develop most frequently in the stomach and are also in the bowel, salivary glands, larynx, and thyroid gland [1]. Unlike the model of gastric MALT lymphoma and *Helicobacter pylori*, no triggering of antigens has been identified in the primary pulmonary MALT lymphoma. It can affect lung parenchyma and bronchi, but only a few references have been made in the literature concerning findings of bronchoscopy, especially when lesions only involve the main bronchus [2,3].

Case Report

A 63-year-old woman who was a housewife in Southern Taiwan complained of intermittent hemoptysis for about one month. The amount was variable from blood-tingled sputum to several milliliters of fresh blood. She was a nonsmoker and was healthy before this event. She had no history of hemoptysis before, although she had one episode of gastric ulcer bleeding two years previous. She did not have fever, chills, body weight loss, epistaxis, epigastralgia or melena accompanied with hemoptysis during this month. Physical examinations produced unremarkable results. Breathing sounds were clear. She had neither peripheral lymphadenopathy nor hepatosplenomegaly. On investigation, hemogram, renal and liver function tests were all within normal limits. There was no abnormal finding from the posteroanterior chest roentgenogram.

She received bronchoscopic examination for prolonged intermittent hemoptysis. An elevated flat plaque was found through a scope. It was located in the anterior wall of the lower trachea just above the carina, and a small portion extended to the left main bronchus. The surface tended to be more white than other normal mucosa, and there were many bleeding spots on the surface (Fig. 1). Bronchoscopic biopsy was performed. The results of histopathology with immunohistochemical analysis consisted of MALT-associated malignant lymphomas.

Then she received computerized tomography examinations of the abdomen and chest. There was no evidence of lymph node involvement, including the thymus, mediastinal nodes, splenic hilar nodes, celiac nodes, portal nodes, paraaortic nodes, iliac nodes and mesenteric nodes. Oral and nasopharynx examinations revealed no
abnormalities. Although she had a previous history of gastric ulcers two years previous, the repeated gastric endoscopy had no significant changes. She also received a bone marrow biopsy from the right posterior iliac crest. The pathology showed no abnormal lymphoid cell infiltration. According to the Ann Arbor staging system, she had stage IE non-Hodgkin’s disease. Therefore, she received radiotherapy over the carina area and upper mediastinum area with 3600 cGy in 180-cGy fractions. Three months later, she received another bronchoscopic examination. The plaque lesion demonstrated a regressive change over the anterior wall of the lower trachea compared with the previous bronchoscopic examination (Fig. 2). The biopsy was performed again, but the pathologic report showed no remnants of the previous tumor. Furthermore, the hemoptysis improved. She received regular follow-up in our out-patient clinic, and there was no evidence of recurrence.

Discussion

Lymphomatous proliferation disorder can involve the lungs as a primary lesion, especially in non-Hodgkin’s lymphomas (NHL). Primary pulmonary lymphoma represents only 0.5-1% of primary pulmonary malignancy and less than 1% of all lymphoma. About 69%-78% of primary pulmonary lymphoma cases have been reported as MALT-type lymphoma [1,2]. The disease often attacks patients from 20-80 years old, but subjects under 30 years old are rarely affected. The incidence in both sexes is equal. Around half of the patients have a history of smoking [2].

About half of the patients with primary pulmonary MALT lymphoma are asymptomatic at presentation, and nearly half of these cases are identified on the basis of abnormal radiological findings by accident. The pulmonary symptoms are nonspecific like cough, dyspnea, chest pain, and occasional hemoptysis, but are more common than constitutional symptoms like body weight loss, fever, night sweats, or fatigue. These symptoms may present for several weeks to months before diagnosis [1-3]. Endobronchial involvement of the disease may cause cough, dyspnea, respiratory distress, atelectasis, obstruction pneumonia, bronchiectasis, or abscess formation due to central airway obstruction. Another form of airway involvement is diffuse submucosa infiltration by tumor cells, and the widespread airway narrowing can cause dyspnea and wheezing, which may be misdiagnosed as asthma [3].

The common findings in chest radiographs of patients with primary pulmonary MALT lymphoma are pulmonary nodules and parenchyma consolidation and/or mass. It may be representative of bilateral or unilateral disease, and the ratio is equal [2, 4]. The lesion is typically 2 to 8 cm in diameter. Up to 50% of cases demonstrate air
bronchograms within the mass or consolidation on the chest radiographs. It is more sensitive to computed tomography. The presence of distended bronchi has the unique radiological appearance of primary pulmonary MALT lymphoma, although the mechanism is not known [1,3]. Other image findings including diffuse alveolar and interstitial infiltrates, atelectasia, pleural effusion constitute around 10% of cases [1-4].

Usually, the diagnosis is difficult to make for primary pulmonary lymphoma because of latent, non-specific clinical presentation and radiographic findings. Although the diagnosis of primary pulmonary MALT lymphoma has been achieved with bronchoalveolar lavage, bronchial biopsy, and transcutaneous biopsy in a few case reports, most cases require thoracoscopic or open lung biopsy [5-7]. Bronchial alveolar larvage fluid appears to be valuable if it shows lymphocytes alveolitis (lymphoid cell count > 20%) [8]. Due to these characteristics of the disorder, the interval between the first clinical or radiological manifestation and diagnosis ranges from 5 months to 8 years [1-3, 8].

Only a few published reports in the literature have described the findings of bronchoscopic examinations of primary pulmonary MALT lymphoma. The most common abnormal bronchoscopic findings of cases have been the inflammatory change of mucosa and bronchial stenosis [1,3,8]. In our case, the bronchoscopic findings only showed elevated plaque in anterior wall of the lower trachea without evidence of tumor infiltration.

There were normal findings from the chest radiographs or computed tomography in our patient. Besides, there was no evidence of parenchyma involvement. The only lesion in this case was the endobronchial plaque-like tumor. In a recent review series of 22 cases, two patients had additional findings of peribronchial thicking in computed tomography, in which they must have had unilateral or bilateral lung parenchyma lesions [2]. It appears there have been no previous cases published under the name of “primary endobronchial MALT lymphoma” via a Medline search.

The outcome of MALT-type primary pulmonary lymphoma is generally favorable. More than 80% of the cases have a five-year survival rate, and the median survival rate has been more than 10 years. The overall survival is better than other types of non-Hodgkin’s lymphoma [1-3]. Clinical features associated with poor prognosis in a series study of primary pulmonary lymphoma included patients over 60 years of age, elevated serum lactate dehydrogenase, elevated serum $\beta_2$ microglobulin levels, Eastern Cooperative Oncology Group performance status of 2 to 4, more than one extranodal site of involvement and failure to enter a complete response after the first phase of therapy [1,3,8].

The choice of treatment in primary pulmonary lymphoma is based on the
histological subtype of the neoplasm, tumor bulk, and comorbid general medical conditions. Antibiotic therapy is inappropriate in primary pulmonary MALT lymphoma, as there is a lack of identifiable chronic antigen stimulation like *Helicobacter pylori* in the model of gastric MALT lymphoma. Surgical resection and radiotherapy are preferred for most localized MALT lymphoma including gastric and pulmonary cases. Oral chlorambucil can be used in patients who have no bulky tumors or who have undergone surgical resection of localized disease [3,8]. For patients with bulky disease, bilateral or extrapulmonary involvement, relapse or progression, treatment options achieving a complete or partial response include single agent regimens with chlorambucil, cyclophosphamide, azathioprine, or steroids or combination systemic regimens such as CHOP (cyclophosphamide, adriamycin, oncovin and prednisone), CVP (cyclophosphamide, vincristine and prednisone). However, combination regimens have not proven more effective than single chemotherapy regimens [1,2]. For our patient, surgery was not indicated because of the lesion located around the carina. We chose local radiotherapy for the localized disease of this patient. Six months later, there was no evidence of recurrence after radiotherapy. Ahmed et al. reported two cases in which radiotherapy was the primary treatment and where one case achieved complete response [2].

MALT lymphoma was the most common type of primary pulmonary lymphoma, although it was still extremely rare in primary pulmonary malignancies. The common findings in chest radiograph were pulmonary nodules and parenchyma consolidation and/or mass. Furthermore, nearly half of the subjects had air bronchograms within the mass in computed tomography. Most of the symptoms were non-specific, and patients were often asymptomatic. The diagnosis of primary pulmonary MALT lymphoma was time-consuming. Our case had an unusual manifestation, which was the involvement of the mainstem bronchus with a normal appearance on radiology. Therefore, primary pulmonary MALT lymphoma should be considered in the differential diagnosis of hemoptysis.

References


Fig. 1. Before radiotherapy, the tumor appeared as an elevated flat plaque found located in the anterior wall of the lower trachea just above the carina. Its surface tended to be whiter than other normal mucosa, and there were many bleeding spots on the surface.
Fig. 2. Following bronchoscopic examination after radiotherapy showing a regressive change in the tumor compared with the previous examination.

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