Case Report
Clinical and pathological characteristics of pulmonary mucosa-associated lymphoid tissue lymphoma: a case report

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Abstract: Introduction: Pulmonary mucosa-associated lymphoid tissue (P-MALT) lymphoma is a relatively rare pulmonary disease. The purpose of our report is to characterize the symptoms, diagnosis and treatment of P-MALT lymphoma. Case report: We reported a case with a history of cough, expectoration, night sweat and multiple lung nodules on chest computer tomography. The pathological analysis of the lung biopsy through VATS demonstrated P-MALT lymphoma. The patient did not take any further treatment. During the following up after the surgery, no changes of symptoms and radiological presentation were detected. Discussions: P-MALT lymphoma is a relatively rare entity and could be easily misdiagnosed. A proper diagnosis could be established by pathological examination on lung biopsy samples. The disease progresses slowly and patients can benefit from chemical and surgical therapy.

Keywords: Pulmonary mucosa-associated lymphoid tissue, lymphoma, pathology characteristics, lymphoepithelial lesions, radiological presentation

Introduction

Pulmonary mucosa-associated lymphoid tissue (P-MALT) lymphoma is an extranodal marginal zone B cell lymphoma. P-MALT lymphoma usually demonstrates a non-specific clinical course. Hence, it is difficult for clinicians to make correct diagnosis. Moreover, the diagnosis and optimal therapy of the disease remain under debate. Even though the diagnosis could be made by comprehensive analysis of medical history, laboratory tests, imaging features and biopsy results, unfortunately, no single of them is specific enough [1, 2]. Therefore, we discuss the histological and clinical characteristics of P-MALT lymphoma that are helpful in identifying this rare entity.

Case report

A 48-year-old man initially had a health check-up in October 2010. Chest computed tomography (CT) scan revealed multiple high density nodules in both lungs (Figure 1A). The patient did not present any symptom of cough, expectoration, chest tightness and shortness of breath, etc. The patient did not smoke and had no history of respiratory illness of tuberculosis or fungal infection. The physical examination revealed no abnormalities. T cell enzyme-linked immune-spot tuberculosis (T-spot) was negative. Then, the patient had not followed up until one year later, he coughed with small amount of white phlegm and presented night sweat intermittently. As there was no change of nodules on CT scan, chronic pulmonary infection was considered and levofloxacin was prescribed for 10 days. His symptoms seemed to be alleviated after taking antibiotics. After that, he did not visit back until January, 2016. He took positive electron transit (PET)-CT scan and it showed a slight increase in the metabolic activity of the nodule in left upper lobe. Maximum standard uptake value (SUV) was 1.6 (Figure 2). The following CT scan in March 2016 showed similar nodules in the left upper lobe and right upper lobe (Figure 1B). For further diagnosis, the patient was qualified for surgical diagnostics and the left upper lobe was wedge resected through the video-assisted thoracoscopic surgery (VATS) lobectomy. The lump...
was yellow-white with size of 30×30×25 mm. The histopathology revealed cystic dilation of alveolus (Figure 3A), centrocyte-like cells, reactive follicles and lymphoepithelial lesions (LELs) with a large amount of lymphocytes around the bronchioles and bronchovascular bundles (Figure 3B), a large number of small lymphocytes accumulated in lung (Figure 3C), uniform eosinophilic material deposition and calcification (Figure 3D). Further immunohistochemical staining showed centrocyte-like lymphocytes positive for CD20 (Figure 3C) and BCL-2, negative for CD3, CD5, CD10, BCL-6 and Cyclin-D1. Ki-67 index was around 5%. Congo red of spe-
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Special staining demonstrated amyloid deposition in orange red (Figure 3D). Fluorescence in situ hybridization (FISH) demonstrated MALT lymphoma positive cells (Figure 4). MALT lymphoma translocation gene 1 (MALT1) gene rearrangement was positive and detected in 30% tumor cells. These results were consistent with the diagnosis of P-MALT lymphoma. Without further treatment, the patient underwent clinical follow-up for 3 months after surgery. No changes in CT scan were detected (Figure 1C).

Discussion

P-MALT lymphoma is a rare disease, which accounts for only 8-10% of all cases of B cell lymphomas and is commonly seen between 50 to 70 years of age [3]. The pathogenesis of this disease has not been clearly elucidated yet. However, it has been reported that inflammatory disorders, smoking, autoimmune diseases such as Sjogren’s syndrome, rheumatoid arthritis, amyloid deposits, collagen vascular diseases, helicobacter pylori infection, acquired immune deficiency syndrome, and pulmonary adenocarcinoma were related to the pathogenesis of MALT lymphoma [4].

The clinical symptoms are usually nonspecific, such as cough, mild dyspnea, chest pain and rarely hemoptysis. The majority of the patients are asymptomatic at the moment of initial diagnosis [5]. Our patient presented the symptoms of cough, expectoration and night sweats intermittently.

The radiographic characteristics of P-MALT lymphoma are various and nonspecific. Besides solitary or multiple pulmonary nodules, masses and consolidation shadow, it was also reported that ground-glass opacity and pleural effusion could be the chest CT manifestations [6]. In the series CT scan of our patient, the CT showed multiple nodules at different sizes scattering in both lungs. The largest one is a calcified nodule with coarse edge in left upper lobe in size of 27×23 mm and the smallest one is a cystic nodule in right upper lobe. With the atypical symptoms, no response to routine antibiotic treatment and nearly unchanged nodules in CT scan around 5 years, it indicated that the lumps are benign or low-grade malignant.

P-MALT lymphoma is difficult to be diagnosed with the clinical symptoms, laboratory examination and CT scan results. To make the diagnosis, biopsy and the following pathology are the only choices. The typical pathology characteristics of MALT lymphoma include small lymphocytes, centrocyte-like lymphocytes, plasmacytoid lymphocytes (Dutcher bodies) and occasional immunoblasts infiltrates. LELs, pleural invasion, giant lamellar bodies, amyloid deposition and reactive follicles are also commonly found. The immunohistochemical profiles of P-MALT lymphoma include positive in CD20 and BCL-2, negative in Cyclin-D1, CD5, CD10 and BCL-6 [2]. Small tissue samples obtained via percutaneous biopsy or TBLB are normally not enough. Therefore, many patients were diagnosed based on the results of surgical biopsies. After VATS resection, the pathology of lung biopsy presented small lymphocytes infiltration, LELs and amyloid deposition in nodules. The immunohistochemical results were
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Positive in CD20 and BCL-2, negative in CD5, Cyclin-D1, BCL-6 and CD10. These results matched major characteristics of MALT lymphoma diagnostic criteria. In hence, P-MALT lymphoma was his final diagnosis.

Currently, the optimal therapy for P-MALT lymphoma remains under debate. The treatment includes simple monitoring, surgery, radiotherapy, chemotherapy, surgery combined with postoperative chemotherapy, chemotherapy followed by radiotherapy. Chemotherapy is still recommended as the primary treatment. CHOP and R-CHOP are the most commonly used chemotherapy regimens. Treatment of P-MALT lymphoma should be based on stage, histology and performance status. The five-year and ten-year survival rates were 90% and 72% respectively [7].

In summary, P-MALT lymphoma is a rare disease that progresses slowly and could be easily
misdiagnosed by its indolent behavior. VATS lung biopsy is an effective method for the diagnosis and treatment of the disease to some extent. Chemotherapy may be an effective treatment.

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Disclosure of conflict of interest

None.

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References


