

A case report of unusual cavity presentation of pulmonary extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue

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Abstract

Rationale: The common CT scan findings of pulmonary MZBL of MALT type include airspace consolidation, nodules and ground-glass opacity. But, to our knowledge, the present case is the first report of a cavity presentation of pulmonary MZBL of MALT type.

Patient concerns: The patient gives his consent and authorizes the photographs featuring his likeness to be published.

Diagnoses: This patient was diagnosed as pulmonary MZBL of MALT type by pathology, immunohistochemistry, and gene rearrangement.

Interventions: The patient was treated with CHOP (cyclophosphamide, adriamycin, vincristine and prednisone) chemotherapy for twice and antibiotics.

Outcomes: He is being followed up for one year, with slight progress in pulmonary MZBL of MALT.

Lessons: This case highlights the need to be suspicious of MZBL of MALT type, when a radiographic image shows cavity lesion. We should consider whether the diagnosis is correct, when the patient's treatment is not effective.

Abbreviations: COPD = chronic obstructive pulmonary disease, DILD = diffuse interstitial lung disease, MALT = mucosa-associated lymphoid tissue, MZBL = extranodal marginal zone B-cell lymphoma, NHL = non-Hodgkin's lymphoma, PCR = polymerase chain reaction.

Keywords: cavity presentation, pulmonary extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue

1. Introduction

We report a case of pulmonary marginal zone B-cell lymphoma (MZBL) of mucosa-associated lymphoid tissue (MALT) type with unusual cavity presentation. Although there are many reports of pulmonary MZBL of MALT type, but cavity presentation of pulmonary MZBL of MALT type has, to our knowledge, never been reported. This case highlights the need to be suspicious of MZBL of MALT type, when a radiographic image shows cavity lesion. The study was approved by the ethics committee review board of the First Affiliated Hospital of Chengdu Medical College. The patient gives his consent and authorizes the photographs featuring his likeness to be published.

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2. Case report

A 63-year-old man presents with a 4-year history of progressive exertional dyspnea and a recurrent productive cough producing white or yellowish sputum. He has a smoking history of 40 pack-years and stopped 4 months ago. He had been admitted to the local hospital many times and received a diagnosis of "COPD with lung abscess." He was treated with antibiotics, which eased the symptoms. A chest CT showed emphysematous changes throughout and a solitary cavity in the left lower lobe (Fig. 1A and B). For the recurring symptom, he was referred to our hospital for the further diagnosis and treatment.

Vital signs were normal, and significant physical examination findings included the following: a barrel chest and the breathing sounds with widely dry rales in lung bilaterally. Significant laboratory findings on admission included the following: WBC count, $4.12 \times 10^9/L$ (48.8% neutrophils and 40.5% lymphocytes); serum tumor makers: CA125 90.56U/mL (normal range <35U/mL), LTA 5.30ng/mL (normal range <3ng/mL). Spirometry showed an FEV₁ of 1.43L (54.3% predicted) and an FEV₁/FVC of 61.4%. Arterial blood gas on room air showed a pH of 7.40, Pco₂ of 48 mm Hg, and Po₂ of 72 mm Hg. Renal function, electrolytes, and coagulation studies were within normal limits. The chest enhanced CT scan is shown in Figure 1C and D.

The patient was started on empiric antibiotic therapy with Ciprofloxacin for yellowish sputum and the cavity of the left lower lobe. Sputum culture showed *Escherichia coli*, and the antibiotics was not changed. The patient underwent fiberoptic bronchoscopy, which revealed no remarkable findings (data not shown). In spite of that, endobronchial biopsies of bronchus in the left lower lobe were obtained and histological examination showed a diffuse infiltrate of lymphocytes (Fig. 2A). Immunocytochemically, the lymphoid cells stained positive for CD20

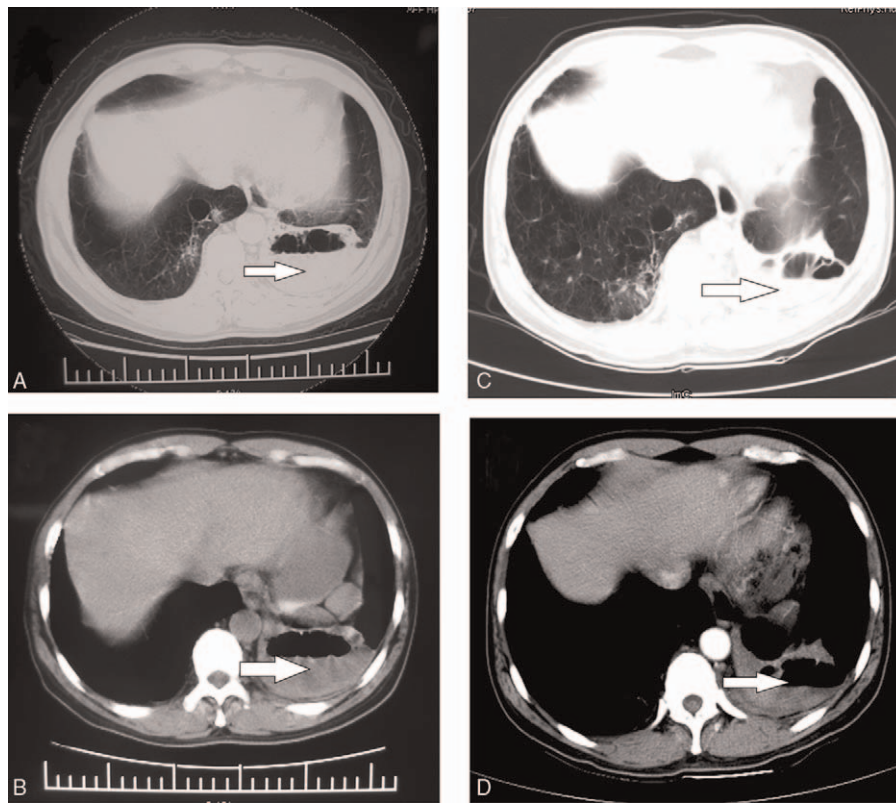


Figure 1. CT scans of the chest. (A) Pulmonary window, (B) mediastinum window: Chest CT scan in the local hospital demonstrated emphysematous changes and a solitary cavity of the left lower lobe. (C) Pulmonary window, (D) mediastinum window: Chest CT scan in our hospital is consistent with the local hospital.

(Fig. 2B) and Ig K (Fig. 2C), but negative for CD21, CD10, CD3, CD23, and CyclinD1 (data not shown). The positive rate of Ki67 was $< 5\%$ (Fig. 2D). Further, the PCR-based analysis showed IGH, IGK gene rearrangement (Fig. 3). Afterward, the patient underwent abdominal CT scan, lymph node of neck color Doppler ultrasound, and bone marrow biopsy, which were negative. But he refused to take esophagogastroduodenoscopy, because he had received the exam which was also negative a few months ago in the local hospital. Finally, the patient was diagnosed COPD with pulmonary MZBL of MALT type and treated with CHOP (cyclophosphamide, adriamycin, vincristine, and prednisone) chemotherapy for twice and antibiotics. He is being followed-up for 1 year, with slight progress in pulmonary MZBL of MALT.

3. Discussion

Primary pulmonary NHLs are making up $< 1\%$ of all NHLs and more than two-thirds of all pulmonary NHLs are MZBL of MALT type.^[1] Its pathogenesis remains unclear, but pulmonary MZBL of MALT type has been associated with chronic inflammatory diseases such as chronic hypersensitivity pneumonitis, diffuse panbronchiolitis, and various autoimmune diseases.^[2,3] Chronic infections also seem responsible for site-specific marginal zone lymphomas, such as with *Helicobacter pylori* for gastric MALT lymphoma, *Campylobacter jejuni* for immunoproliferative small intestine disease, *Borrelia burgdorferi* for B-cell cutaneous lymphoma, and *Chlamydiae psittaci* for ocular adnexal lymphoma. Moreover, other infections, such as *Mycobacterium tuberculosis*^[4] and *Chlamydiae*^[5] are involved

pulmonary MZBL of MALT type. But the specific infection for the development of malignancy still remains unknown.

Previously studies have demonstrated that the median age of pulmonary MALT lymphoma are > 50 years old.^[2,3] Some patients are asymptomatic, some have nonspecific respiratory symptoms, such as cough, sputum, exertional dyspnea, and the others have system symptoms including fever, fatigue, and weight loss.^[6] Physical examination is usually noncontributory.

The chest radiographic images of the disease are diverse, including consolidations, nodules, bronchiectasis, DILD. Bae et al classified the pattern of parenchymal lesions into the following 4 different patterns including: single-nodular or consolidative; multiple nodular or areas of consolidation; bronchiectasis and bronchiolitis; and DILD.^[7,8] But the cavity presentation has not yet been described. We are wondering whether it is a new pattern which we have ignored. The mechanism of the cavity formation in pulmonary MZBL of MALT type remains unknown, but it is thought to be due to a check-valve mechanism like the other lung cancer.^[8] Another reason could be a result of avascular necrosis and destruction of the pulmonary alveoli by excessive mucus.^[9] Furthermore, we hypothesize repeated infection might be one of the reasons. Cavity lesions are nonspecific and may be seen in various lung diseases, such as lung abscess, tuberculosis, aspergillus, Wegener's granulomatosis, and pulmonary sequestration. Then how to differentiate the primary pulmonary MZBL of MALT type from other lung diseases with cavity lesions. It mainly depends on the results of pathology, immunohistochemistry, and gene rearrangement. Furthermore, a review of the literature revealed this case to be anecdotal as it is extremely unique for a primary

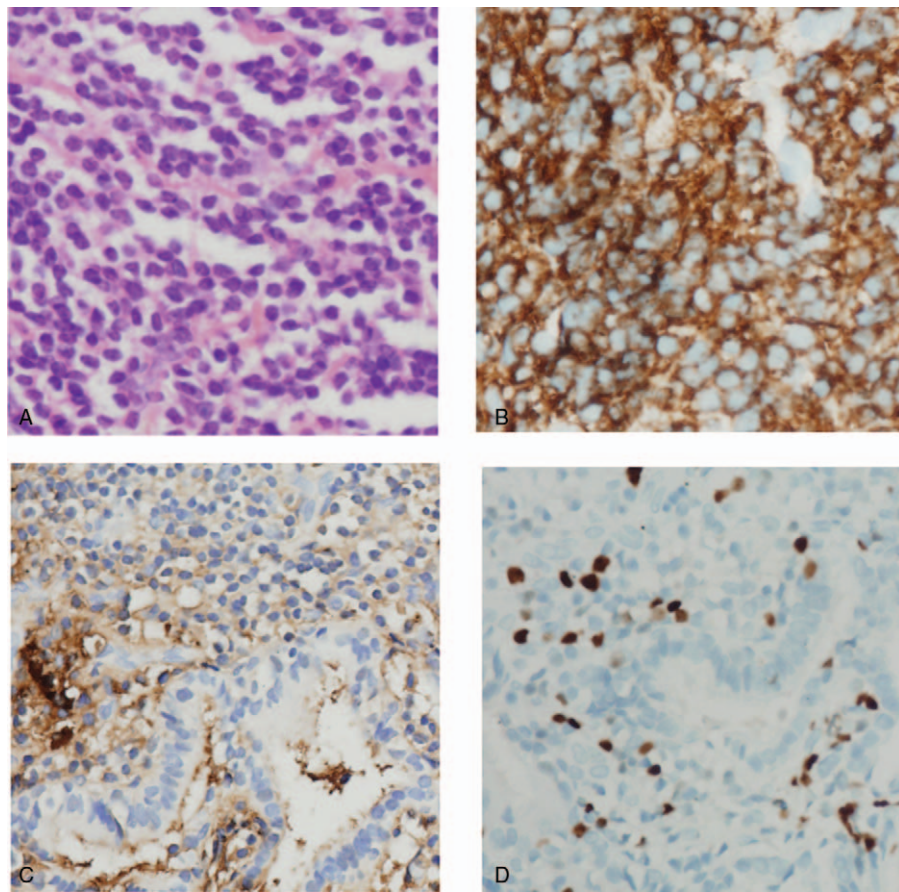


Figure 2. Morphology and immunohistochemistry. (A) Endobronchial biopsy specimen shows a diffuse infiltrate of lymphocytes (hematoxylin–eosin, original magnification $\times 300$). (B) Endobronchial biopsy specimen shows the lymphoid cells in cystic walls stained immunohistochemical positive for CD20 (immunoperoxidase, original magnification $\times 300$). (C) Endobronchial biopsy specimen shows the lymphoid cells in cystic walls stained immunohistochemical positive for IgK (immunoperoxidase, original magnification $\times 300$). (D) Endobronchial biopsy specimen shows the positive rate of Ki67 was $<5\%$ (immunoperoxidase, original magnification $\times 300$).

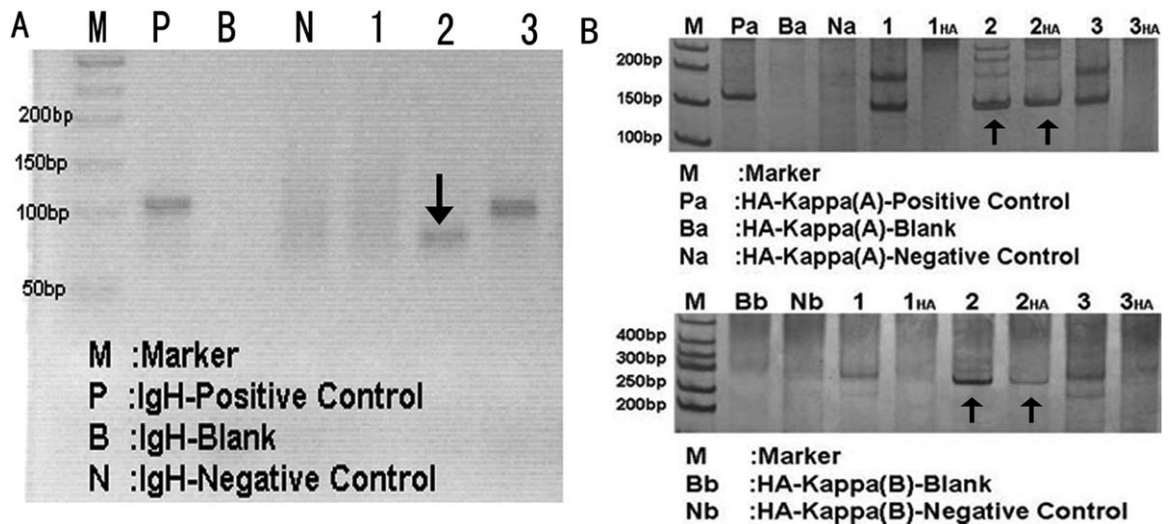


Figure 3. The gene rearrangement of endobronchial biopsy specimen. (A) Clonal rearrangement band of IgH is detected by PCR analysis (Arrow). (B) Clonal rearrangement band of IgK is detected by PCR analysis (Arrow). Lanes 1 and 3 were specimens of other cases, which were detected simultaneously with that from the targeted patient discussed in the text.

pulmonary MZBL of MALT type to present in the form of cavity. This case leads us to the conclusion that when a radiographic image shows cavity presentation, the possibility of pulmonary MZBL of MALT type has to be considered in the differential diagnosis.

Pulmonary MZBL of MALT is a rare disease, and no standard treatments have been defined yet. Currently, treatment of pulmonary MZBL of MALT type includes surgery, radiotherapy, and chemotherapy. But study found that patients who received chemotherapy had superior progression-free survival (PFS) than those treated with upfront surgery.^[10] So our patient was treated with chemotherapy. But the patient was only treated with CHOP (cyclophosphamide, adriamycin, vincristine, and prednisone) chemotherapy for twice and abandoned chemotherapy due to side effects. That is the reason why his disease made slight progress. In our opinion, chemotherapy should be considered as first-line options for the treatment of pulmonary MALT lymphoma, and for selected patients, watch-and-wait should be another rational option.

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Author contributions

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