Case Report

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Primary pulmonary mucosa-associated lymphoid tissue lymphoma with extensive lung involvement and negative autoimmune and inflammatory background: A case report and literature review

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Abstract:

Primary pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma is a very rare presentation of MALT lymphoma. The presence of a completely negative autoimmune and inflammatory background makes it a real challenge and very rare presentation (probably the second reported case in the literature). We report a case of primary pulmonary MALT lymphoma with negative autoimmune background, demonstrating as multifocal bulky variceal masses causing significant clinical symptoms.

Keywords:

Extranodal marginal zone lymphoma, negative autoimmune mucosa-associated lymphoid tissue lymphoma, primary pulmonary mucosa-associated lymphoid tissue lymphoma

Introduction

Extranodal marginal zone lymphoma (ENMZL) of mucosa-associated lymphoid tissue (MALT) is a lymphoproliferative disorder that morphologically comprises small heterogeneous B lymphocytes developing in an indolent pathway. It represents 7% of all non-Hodgkin B cell lymphomas, occurring at the average age of diagnosis in the sixth decade of life. ENMZL emerges as a result of immune cross-reaction provoked by persistent exposure to inflammatory or autoimmune stimuli. The most established example of which is the causative effect of *Helicobacter pylori* in 92% of stomach MALT lymphomas.[1,2]

Although the stomach is the most affected organ, it can arise in other organs such as salivary glands, skin, orbits and conjunctivae, thyroid, breast, liver, and lungs.[1] Lung as the primary site of MALT lymphoma, whether parenchymal or bronchial tissue, is even more rare, representing less than 1% of all lung malignancies, and only 3%–4% of all ENMZL.[3]

The prospect of acquiring lung lymphoma was explained in the literature as persistent exposure to inflammatory or autoimmune process. Examination of the basis of the pathophysiology revealed that the B cell clone has a B cell receptor with rheumatoid factor (RF) activity exposed to the Fc part of Immunoglobulin-G. Subsequently, it was

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concluded that the indicated polyclonal B-cells triggered in the autoimmune inflammatory setting achieve extra gene mutations that give rise to B cell lymphoma.[2]

Case Report

A 52-year-old Saudi female patient, nonsmoker, not known to have any chronic medical illness presented with significant nonintentional weight loss of 20 kg in 2 years, progressive shortness of breath, and right pleuritic chest pain of three months duration. There was no other relevant past medical or family history. She had no history of environmental exposures (industrial contaminants), contact with ill patients and had unremarkable family history for any autoimmune diseases or malignancies.

She presented to the hospital in respiratory distress with evidence of desaturation to 85% at room air. Physical examination was positive for decreased air entry in the right lower lobe, with coarse crepitations observed in the right middle and left upper lobes.

Her lab works showed normal blood count with differentials, normal liver function test, C-reactive protein of 0.8 mg/dL, Procalcitonin <0.02 ng/mL, erythrocyte sedimentation rate of 106 mm/h. All septic workups from sputum and blood were negative for bacterial, viral, and fungal microorganisms. Antinuclear antibody was 160 CU, Anti-double Stranded DNA <1:10, antineutrophil cytoplasmic antibodies <20 CU, Anti-Sjogren's-Syndrome-Related-Antigen A <4.9 CU, Anti-Sjogren's-Syndrome-Related-Antigen B <3 CU, RF <20, Myeloperoxidase <3.2 CU, Proteinase 3 (PR3) <2.3 CU, Anti-Cyclic Citrullinated peptide <4.6 CU, Anti Smith <3.3 CU, Anti-Jo1 <2.2 CU, Anti-ribonucleoprotein <3.5 CU, 34 Anti-SCL70 <1.2 CU and all other autoimmune workup were negative.

Plane chest X-ray with posterior-anterior view showed multifocal consolidation with right pleural effusion. Computed tomography scan revealed a large mass-like consolidation occupying the right middle lobe bulging into the horizontal and oblique fissures with significant mass effect upon the distal superior vena cava and the right atrium, and additional multifocal consolidations with air bronchograms and surrounding ground-glass opacities involving all other segments [Figure 1].

Owing to the lack of response to multiple antibiotic courses, bronchoscopy was performed, and showed multiple bronchial masses that were variceal-like and friable to touch [Figure 2]. Bronchial alveolar lavage was taken, and workup including cultures, gram stains, acid-fast bacillus stain and culture were sent, all of which were negative.

Interventional radiology team were involved later for direct tissue sampling. Histopathology reported microscopic picture of diffuse, uniform monotonous proliferation of small, round blue cells with distinct cell borders, nuclei with regular contours, inconspicuous nucleoli and abundant pale cytoplasm. Mitotic figures were seen. There was mild-to-moderate plasma cell infiltration in the background. Immunohistochemistry study was performed. The tumor cells were positive for CD45, CD20, and CD38. Ki-67 (30%–40%), but CD5, CD23, CD10, Synaptophysin and Cyclin D-1 were all negative. Bone marrow showed no morphological or immunophenotypic evidence of marrow infiltration by lymphoma with additional interstitial scattered positivity to CD79a and CD3.

Positron emission tomography (PET) scan reported multifocal large fluorodeoxyglucose (FDG) avid consolidations in the lungs involving the anterior segment of the left upper lobe measuring 6.7 cm × 10 cm (SUVmax 6.4), left lower lobe measuring 11.3 cm × 6.3 cm (SUVmax 6) and right middle lobe measuring 9.9 cm × 14.5 cm (SUVmax 6.4) with lymphomatous involvement associated with bilateral pleural effusion and additional upper abdominal large retroperitoneal FDG avid lymph node at the aorto-caval region measuring 20 mm in short axis with SUVmax 6.9.

The patient was counseled and agreed to receive rituximab 375 mg/m^2 and bendamustine 90 mg/m^2 protocol, by the time of the write-up of this paper, three cycles were completed with good tolerance and an improvement in her overall clinical condition (clear weight gain and complete recovery of clinical complaints) and proven PET response.

Discussion

MALT is a lymphoproliferative disorder morphologically comprising small heterogeneous B lymphocytes that develop in an indolent pathway. While recent data support the traditional association with chronic persistent inflammatory and autoimmune processes,[4] diagnosis of this case was a real challenge because of its negative profiles.

The acute course of the disease which resembled the more aggressive type of lymphoma (Diffuse Large B cell) went against the nature of MALT lymphoma indolence in which most cases are usually kept under observation for years without treatment (wait and watch approach). The variceal appearance of the mass lesion on bronchoscopy was unique and distinct from the reported cases in the literature [Figure 2].

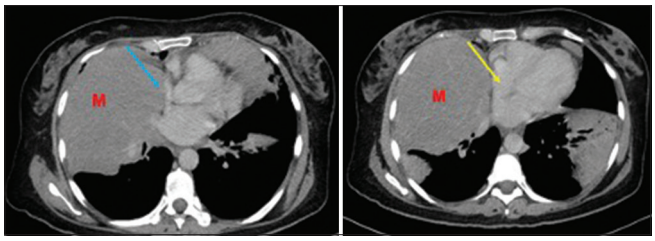

Figure 1: Large mass-like consolidation occupying the right middle lobe with significant mass effect upon the distal superior vena cava and the right atrium (RA). (M) The mass (blue arrow) distal superior vena cava (yellow arrow) RA

Conclusion

Physicians may miss the diagnosis of the earlier stages of MALT lymphoma without further investigation because of the classic benign behavior and background of the disease. This case raises awareness of atypical presentation of MALT lymphoma (the unusual lesion presentation and the complete absence of autoimmune and inflammatory background with negative smoking history or any predisposing factor).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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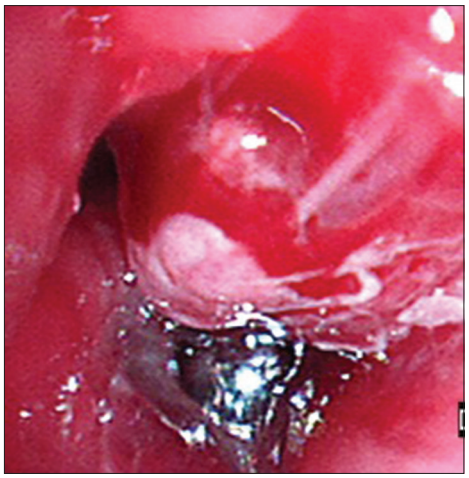

Figure 2: The variceal-like mass in the bronchoscopy

Conflicts of interest

There are no conflicts of interest.

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