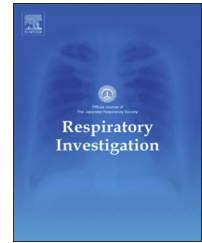




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Short Communication

Primary Sjögren's syndrome with Waldenström's macroglobulinemia presenting as unilateral bloody pleural effusion

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1. Introduction

Approximately 1.5–9% of patients with Sjögren's syndrome (SjS) have pulmonary manifestations, but pleuritis causing pleural effusion is a rare complication [1]. However, it is well known that patients with primary SjS may have combined

hematologic disorders such as anemia, Waldenström's macroglobulinemia (WM), and non-Hodgkin's lymphoma [2]. WM can also result in pulmonary complications, and has an incidence of 3–5% [3]. Winterbauer reviewed the findings of pleural fluid analyses from five WM patients with pleural effusion and reported that the effusion was hazy and amber

Abbreviations: CPA, cyclophosphamide; LCA, leukocyte common antigen; PSL, prednisolone; sIL-2R, soluble interleukin-2 receptor; SjS, Sjögren's syndrome; WM, Waldenström's macroglobulinemia.

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in color [4]. Bloody pleural effusion is a very rare manifestation of SjS or WM. This is the first report describing a case of primary SjS with WM presenting as bloody pleural effusion.

2. Case report

A 61-year-old woman was admitted to our hospital due to cough and dyspnea. She complained of dry cough without chest pain, which was aggravated in the right lateral decubitus position. A chest radiogram showed right-sided pleural effusion. The anti-nuclear antibody titer on hospital admission was 1:320 and the anti-SS-A and anti-SS-B titers were 96.5 and 122.0 U/mL (normal levels: < 15 and < 25 U/mL, respectively). Tumor markers were negative, with the exception of the soluble interleukin-2 receptor (sIL-2R; 3054 U/mL). There was no evidence of infection, malignancy, or collagen diseases other than SjS. The pleural fluid was exudative and had a red, wine-like color as shown in Fig. 1A. Anti-SS-A/SS-B antibodies and IgM levels were remarkably increased and had a high level of lymphocytes (76%). Frequent cytologic examination of the pleural fluid did not show any malignant cells. An ophthalmologic examination included a Schirmer test that was positive; a lip biopsy showed mononuclear leukocyte permeation around the salivary gland ducts. Based on these results, the patient was diagnosed with primary SjS. However, serum protein electrophoresis showed an M-peak corresponding to γ -globulin (Fig. 1B), and serum immunoelectrophoresis revealed M-bow formation with IgM κ . Bone

marrow examination revealed the presence of lymphoplasmacytic cells (Fig. 1C), and flow cytometry of the bone marrow also showed monoclonal proliferation of B cells presenting IgM κ , corresponding to the high serum level of IgM. This confirmed the diagnosis of WM according to diagnostic criteria [5]. Accordingly, the case was diagnosed as primary SjS with WM.

First, the patient was treated with cyclophosphamide (CPA) for the WM, because she had an embolism on the fundus of the eye due to hyperviscosity syndrome. However, the bloody pleural effusion was not regulated despite the decrease in serum IgM levels (Fig. 1D). We could not exclude the possibility of malignancy, as SjS complicated with a bloody pleural effusion has never before been reported. Therefore, flexible thoracic endoscopy was performed under local anesthesia to obtain pleural tissues. There were no remarkable findings on either the parietal or the visceral pleura. A pleural biopsy showed lymphocyte infiltration of the pleural tissues, which might be the cause of pleural effusion secondary to SjS; however, the result was not a specific finding of SjS. The tissue was positive for surface markers of both T cells and B cells (Fig. 1E); no malignant cells were detected. There was no monoclonality of the lymphocytes based on flow cytometric analysis. These results did not correspond to the findings of malignant lymphoma or lymphocytic lymphoma.

As the patient's leukocyte count decreased to 1000/ μ L, the CPA was discontinued and corticosteroid therapy was initiated. The bloody pleural effusion began to abate 10 days

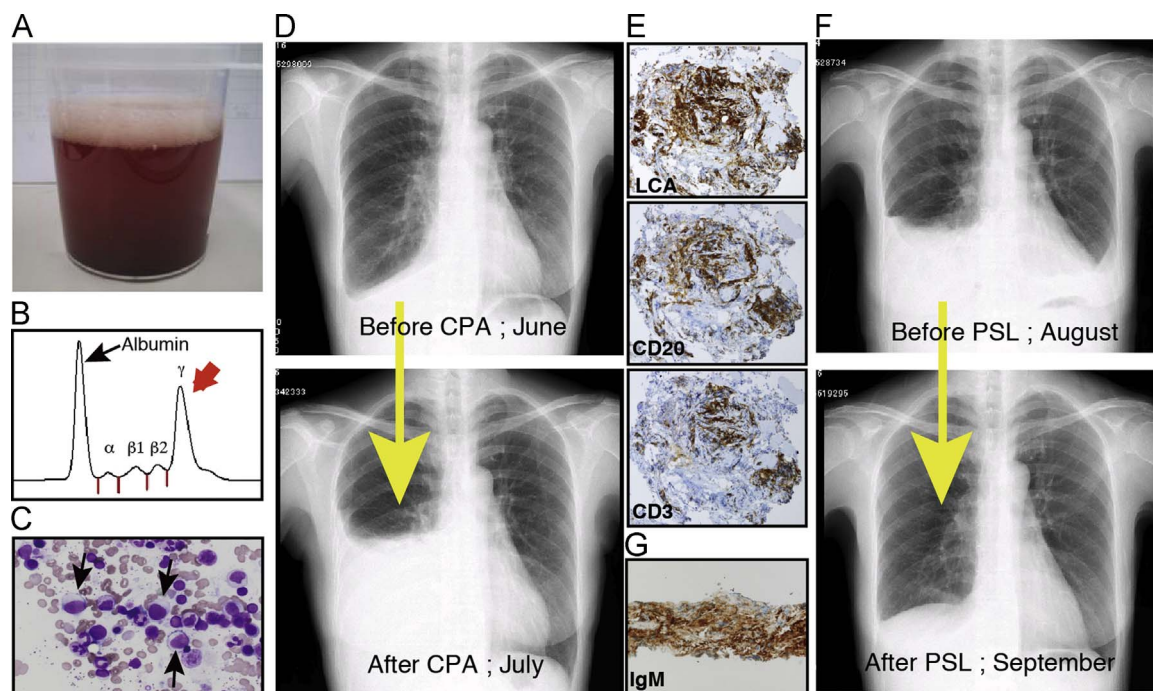


Fig. 1 – Serum electrophoresis, bone marrow analyses, pleural immunohistochemistry, and clinical course of right-sided pleural effusion. A) Bloody pleural effusions in the right lung. B) Serum protein electrophoresis showing an M-peak of γ -globulin (red arrowheads). C) Lymphoplasmacytic cells in a bone marrow examination. Lymphoplasmacytic cells (black arrowheads) in the bone marrow with normal lymphocytes. D) Worsening pleural effusion despite CPA therapy. E) Immunohistochemistry of right pleura with leukocyte common antigen (LCA), CD20, and CD3 antibodies. LCA-positive cells stained positive for CD20 or CD3. F) Decreased pleural effusion with PSL therapy. G) Immunohistochemistry of pleural tissue with IgM antibodies.

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