

Pleuritis associated with primary Sjogren syndrome

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Keywords

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Introduction

Sjogren's syndrome (SjS) is a systemic autoimmune disease characterized by sicca symptoms. Many organic changes have been reported in patients with SjS. Interstitial pulmonary fibrosis and tracheobronchial sicca are the most common presentation of pulmonary involvement in primary SjS [1]. However, it is rarely accompanied by serositis such as pleuritis or pericarditis. We herein present a case of pleuritis complicated with primary SjS diagnosed with infiltration of lymphocytes on pleura by thoracoscopic pleural biopsy.

Case Report

A 71-year-old woman had an 8-year history of SjS, which was treated with 2 mg of daily oral prednisolone. In 2011, left pleural effusion was incidentally found on chest radiograph. Because she had no respiratory symptoms, a careful observation was provided without any additional treatment. In September 2015, she developed dyspnea on exertion, and chest computed tomography (CT) revealed increased bilateral pleural effusion and newly developed ground glass opacity in the right lung field (Fig. 1B).

Abstract

We herein present a case of a 71-year-old woman with primary Sjogren's syndrome (SjS), who developed bilateral pleural effusion and ground glass opacity during treatment with low-dose prednisolone. The pleural effusion and bronchoalveolar lavage fluid revealed elevation of lymphocytes. Thoracoscopic pleural biopsy showed infiltration of lymphocytes with no evidence of other diseases, confirming SjS-related pleuritis. Therefore, we initiated 20 mg prednisolone and pleural effusion was rapidly resolved. Our results indicate that SjS can be rarely complicated with pleuritis. In addition, thoracoscopic pleural biopsy and a rapid response to steroid treatment would be helpful for diagnosing SjS-related pleuritis.

A physical examination revealed reduced air entry on the left side, dry mouth, and dry eyes. Raynaud's phenomenon and eruptions or swelling of any joints were not observed. The results of a complete blood count, urinalysis, biochemical studies, and a coagulation test were normal. The level of brain natriuretic peptide was 57.1 pg/mL (normal < 18.4). The anti-nuclear antibody titre was ×40 with a homogeneous and speckled pattern, while the rheumatoid factor level was 3.0 IU/mL (normal < 15.0). The patient was negative for anti-cyclic citrullinated peptide antibodies, anti SS-A/Ro antibodies and SS-B/La antibodies.

The pleural effusion was turbid and exudate. The pleural fluid total leukocyte count was 1420/μL (neutrophils 5%, lymphocytes 95%). The adenosine deaminase level was normal. No malignant cells were detected in the effusion. Bronchoalveolar lavage fluid (BALF) from the right middle lobe bronchus showed an elevated level of lymphocytes (59%), and the CD4/CD8 ratio was 0.63. Bronchoalveolar lavage fluid and pleural effusion samples were negative for acid-fast bacteria. Because transbronchial lung biopsy did not provide any specific findings, medical thoracoscopy under local anaesthesia was performed. No abnormal endoscopic findings were seen in the left pleural cavity

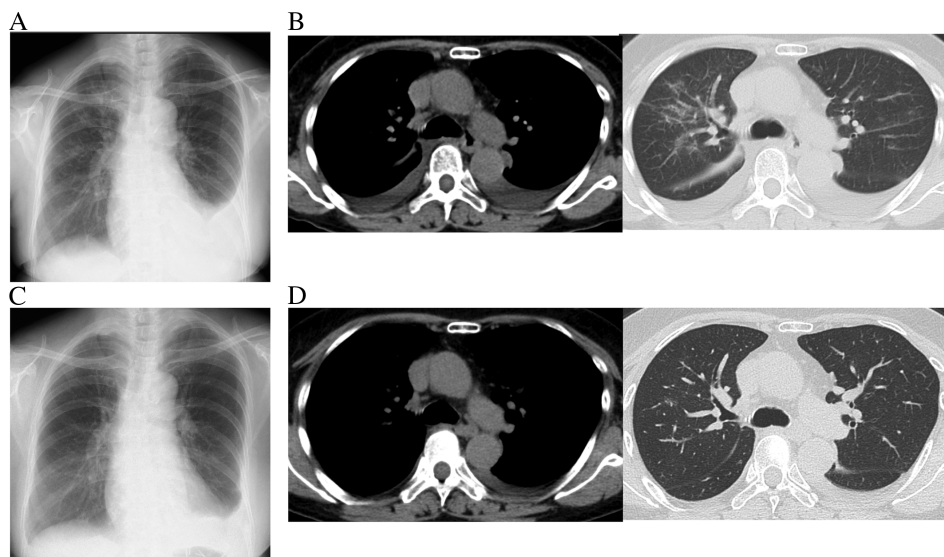


Figure 1. (A) A chest radiograph reveals diffuse ground glass opacity in the right lung fields and left-sided effusion. (B) Chest computed tomography images at the tracheal carina level show ground glass opacity in the right lung fields and bilateral pleural effusion. (C, D) A chest radiograph and chest computed tomography images after steroid therapy. Bilateral pleural effusion and ground glass opacity are decreased.

(Fig. 2A). Biopsy revealed a marked infiltration of lymphocytes, with no evidence of lymphoma or malignant mesothelioma (Fig. 2B). All of the cultures on effusion, BALF, and biopsy specimens were negative for acid-fast bacilli and fungi. Based on the pathological findings, the diagnosis of SjS-related pleuritis was confirmed. Therefore, prednisolone at 25 mg was initiated, and the pleural effusion and ground glass opacity improved immediately, as shown in Figure 1C, D.

Discussion

We herein reported a case of SjS that presented with pleural effusion during the clinical course. Our results provide two clinical implications, described below.

First, pleural effusion can rarely develop in primary SjS patients during the clinical course. In general, pleuritis is a rare complication in primary SjS patients. In fact, a previous study reported that no pleural effusion was observed in 40 cases of primary SjS, although it was observed in 2 of 26 cases (8%) of secondary SjS [2]. To our knowledge,

there have been 11 reported cases of primary SjS complicated by pleuritis [3–13] (Table 1). Among them, only three patients developed pleural effusion during the clinical course, whereas the remaining eight were diagnosed simultaneously with primary SjS and pleural effusion. Of these eight cases diagnosed simultaneously, six had subclinical SjS, which lacked xerostomia. In the present case, pleural effusion emerged during the clinical course of treatment. Taken together, the present and previous findings indicate that pleural effusion can develop in primary SjS patients, regardless of whether or not the patient has undergone prednisolone treatment.

Second, thoracoscopic pleural biopsy and a rapid response to steroid treatment are helpful for diagnosing SjS-related pleuritis. The pathological findings of primary SjS-associated pleuritis are still unclear. In the present case, lymphocytes remarkably infiltrated the pleura with no evidence of malignancy or tuberculosis. These findings seemed non-specific but were strongly suggestive of an autoimmune reaction. Indeed, the pathological findings on pleural biopsy performed in six patients with primary SjS-

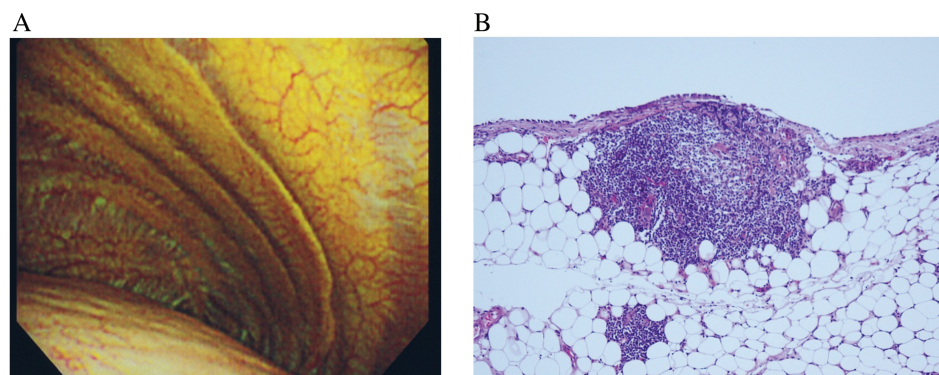


Figure 2. (A) The macroscopic appearance of the parietal pleura during thoracoscopy. No abnormal findings are seen on the left pleura. (B) A pleural biopsy specimen shows infiltration of lymphocytes into the pleura (Haematoxylin and eosin stain).

Table 1. Reported cases of pleural effusion associated with primary Sjogren's syndrome.

Reference	Reported year	Age	Gender	Symptoms	Pleural biopsy	Steroid treatment	Timing of pleuritis and SjS
3	1989	64	F	Chest pain	—	—	During the clinical course of SjS
4	1995	40	F	Fever	—	Prednisolone 60 mg	Simultaneously
5	1995	62	M	Fever	Done	Prednisolone 40 mg	Simultaneously
6	1996	53	F	Cough	—	Steroid pulse therapy	During the clinical course of SjS
7	1997	70	M	Cough	Done	Prednisolone 30 mg	Simultaneously
8	2000	45	F	Fever	—	Prednisolone 40 mg	Simultaneously
9	2000	73	M	Dyspnea	Done	Prednisolone 30 mg	Simultaneously
10	2008	65	M	Cough, dyspnea	Done	Prednisolone 30 mg	Simultaneously
11	2012	63	M	Cough, dyspnea, chest pain	—	Prednisolone 40 mg	Simultaneously
12	2015	42	F	Chest pain	Done	—	Simultaneously
13	2015	69	F	Cough, sputum, chest pain	Done	Methylprednisolone 120 mg	During the clinical course of SjS

F, female; M, male; SjS, Sjogren's syndrome.

In all cases, the pleural effusion showed lymphocyte-dominant cytology.

associated pleuritis were identical to those of our present patient. In general, pleural biopsy via thoracoscopy is considered essential for obtaining important information for diagnosing malignant pleural effusions and tuberculous pleurisy when pleural fluid cytology provides no significant findings. In malignant pleural effusions, the diagnostic rate of pleural fluid cytology remains only 62% [14], and therefore, thoroscopic pleural biopsy is sometimes necessary. Although the pathological findings in primary SjS-related pleuritis are non-specific, thoroscopic pleural biopsy should be considered in order to confirm the diagnosis. However, it is sometimes difficult to perform thoroscopic biopsy, especially in patients with poor performance status. We believe that a rapid response to steroid treatment, which was observed in the present case, will also be helpful in diagnosing SjS-related pleuritis. Although the optimum treatment has not yet been established, eight of nine previous reports cited revealed a rapid improvement of pleuritis on treatment with steroids. The pathological findings of lymphocyte-abundant BAL and lymphocyte-infiltration of pleura support the efficacy of steroid therapy. Thus, in SjS patients with pleural effusion of unknown cause, a rapid response to steroids is also helpful for diagnosing SjS-associated pleuritis; however, the possibility of *Mycobacterium* infection should be excluded before the administration of steroids.

In conclusion, we herein reported a case of primary SjS complicated with pleuritis. Chest physicians should be aware that pleural effusion can develop during the clinical course in primary SjS patients. In addition, thoroscopic

pleural biopsy and a rapid improvement with steroid treatment are helpful for diagnosing SjS-related pleuritis.

Disclosure Statements

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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