Churg-Strauss Syndrome with Pleural Involvement

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Abstract

A 51-year-old Japanese man with Churg-Strauss Syndrome (CSS) diagnosed by pleural biopsy is described. He was hospitalized because of high fever and bilateral knee, elbow and shoulder joint pain. Chest roentgenogram and chest computed tomography (CT) scan revealed bilateral massive pleural effusion. Pleural biopsy revealed eosinophilic infiltration and necrotizing granulomas. He was treated with oral prednisolone and his symptoms improved. This is the first report of CSS diagnosed by pleural biopsy. (Internal Medicine 39: 976–978, 2000)

Key words: allergic granulomatous angiitis, bronchial asthma, pleural biopsy, pleuritis

Introduction

Churg and Strauss (1) proposed allergic granulomatosis and angiitis as a disease to be discriminated from classical periarteritis nodosa (PN) based on its clinicopathological feature of necrotizing vasculitis occurring exclusively in asthmatic patients. Therefore, allergic granulomatosis and angiitis is also referred to as Churg-Strauss Syndrome (CSS), and is characterized by hypereosinophilia, bronchial asthma, and vasculitis. Histological findings of CSS observed in the heart, liver, spleen, lungs, skin (2) and sural nerves (3) are well recognized. Although CSS with pleural involvement has been encountered, this is the first report of CSS diagnosed by pleural biopsy.

Case Report

A 51-year-old Japanese man was admitted to our hospital complaining of high fever, dyspnea, and bilateral knee, elbow, and shoulder joint pain in August 1999. He had been diagnosed with bronchial asthma at the age of 39 years and was instructed to take 400 mg/day of oral theophylline. No specific family history was identified. He had worked as an office worker for about 30 years and had never had occupational exposures.

Physical examination on admission revealed an acutely ill looking man; his temperature was 37.5°C, heart rate was 69 beats/min, respiratory rate of 26/min, and blood pressure of 118/70 mmHg. No lymph nodes were palpable. Neurological examination revealed loss of the patellar tendon reflexes. Multifocal sensory loss was present. He had no abnormal findings including eruptions or purpura in the skin.

Laboratory investigation demonstrated mild anemia, inflammatory reaction, and marked eosinophilia: erythrocyte count, $385 \times 10^4/\mu\text{l}$, hemoglobin, 11.8 g/dl; leucocyte count, 14,500/ μl (neutrophils, 32.3%, eosinophils, 49.5% (normal range [NR] 0.2–6.8%), basophils, 0.4%, monocytes, 3.2%, and lymphocytes, 14.6%), erythrocyte sedimentation rate (ESR), 96 mm/h. The hepatic and biliary enzymes such as aspartate aminotransferase (AST), alanine aminotransferase (ALT), lactate dehydrogenase (LDH), alkaline phosphatase (ALP), leucine amino peptidase (LAP), and γ -glutamyltranspeptidase (γ -GTP) were normal. On renal function tests, blood urea nitrogen (BUN) and creatinine levels were normal. C reactive protein was 6.6 mg/dl (NR 0 mg/dl–0.5 mg/dl). The immunoglobulins were as follows: IgG, 1,210 mg/dl; IgA, 371 mg/dl; and IgM, 160 mg/dl; IgE, 1,700 U/ml (NR <250 U/ml). Tests for p-ANCA, c-ANCA, anti-nuclear antibody, and anti-DNA antibody were negative. A test for rheumatoid factor was positive. The CH50 and C3 and C4 complement proteins were normal. Urinary protein and sugar were negative, and the urine sediment showed no red blood cells. Pulmonary function tests demonstrated respiratory impairment (%VC, 64.6%; FEV1.0%, 58.9%). Chest roentgenogram (Fig. 1) and chest computed tomography (CT) scan revealed bilateral massive pleural effusion. As revealed by pleurocentesis, the nature of the pleural effusion included the specific gravity of 1.03, protein of 4.0 g/dl, and marked eosinophils. A provisional diagnosis of CSS with massive pleural effusion due to pleuritis was made, based on these imaging and immunological examinations, and pleural biopsy was therefore performed. On pleural biopsy, pleural thickening, infiltration by eosinophils (Fig. 2A), and necrotizing granulomas were noted (Fig. 2B).

The patient was treated with oral prednisolone (PSL) 60 mg/day. The fever dissipated quickly. The patient's dyspnea and arthralgia were alleviated after 2 weeks' administration of PSL.

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Figure 1. Chest roentgenogram on admission shows bilateral pleural effusion.

and the pleural effusion disappeared. Sural nerve biopsy after steroid therapy showed decreased myelinated fibers and axonal degeneration (Fig. 3).

Discussion

CSS is sometimes called allergic granulomatous angiitis. This disease which was distinguished from PN by pathologists Churg and Strauss in 1951 (1), is characterized by hyper-eosinophilia and systematic vasculitis occurring in individuals with bronchial asthma (1, 4). There are reports of death from CSS (3) or of poor prognoses due to perforation of the intestine (5), organ failure (6–10), and damage to the CNS (4, 7). In most cases, systemic vasculitis occurs within 3 years after the onset of bronchial asthma (11). In one report it occurred 14 years after the onset of bronchial asthma (12). The present case broke out 12 years after the onset of bronchial asthma, and clinical features were hypereosinophilia, arthralgia, pleural effusion, and multiple mononeuropathies.

The incidence is thought to be 1 in 5,000 asthma patients, and the type of bronchial asthma involved is usually the atopic type (4) or the adult type (11). A review of the literature revealed mononeuritis multiplex (85%), cutaneous involvement (57%), gastrointestinal involvement (50%), cardiac involvement (42%), and pulmonary involvement (28%) in patients with CSS (11). As in the present case, pleural effusions are not uncommon

A

B

Figure 2. Pleural biopsy revealed (A) thickening of the pleura and infiltration of eosinophils (HE stain, ×40) and (B) necrotizing granulomas (HE stain, ×100).

mon (29%) and typically contain large numbers of eosinophils (4, 6).

The classic histological findings of CSS (tissue infiltration by eosinophils, extravascular granulomas and necrotizing vasculitis) may be sparse or widely distributed, and have been encountered in most organs. Granulomas are most commonly observed in the heart, liver, spleen, lungs and skin (2). Nerve biopsies in CSS show decreased myelinated fibers and axonal degeneration (3). These biopsies from heart, liver, spleen, lung, skin, and peripheral nerves may be missed. Thus, pleural biopsy is taken into consideration as a diagnostic modality when patients have abnormal findings suggesting pleuritis such as pleural effusion or pleural thickening, as in the present case. There have been no previously reported cases of CSS with abnormal pleural findings, although pulmonary infiltrates or pleural effusion are common clinical features (4). We found that pleural biopsy was useful for making a diagnosis of CSS.

In conclusion, we reported a case of CSS diagnosed 12 years

A

B

Figure 3. Sural nerve biopsy (A) (HE stain, $\times 80$) and teased-fiber specimen (B) ($\times 63$) show decreased myelinated fibers and axonal degeneration.

after the onset of bronchial asthma. There have been no previous case reports of CSS with abnormal pleural findings that were diagnosed by pleural biopsy.

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