Fulminant Myocarditis in Polymyositis

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Cardiac involvement in patients with polymyositis is usually asymptomatic and associated with a mild clinical course. A female patient with muscle weakness and cardiogenic shock, who was diagnosed with polymyositis and fulminant myocarditis, is described. A large amount of methylprednisolone, in addition to intra-aortic balloon pumping and percutaneous cardiopulmonary support, led to the recovery of her cardiac function. However, a massive cerebral embolism occurred and she died. Postmortem histopathological examination showed necroses of muscles and diffuse invasion of mononuclear cells in both the myocardium and the biceps muscle of her arm. Although the mechanism of cardiac dysfunction is not clear, immunosuppressive therapy was effective for fulminant myocarditis in the present case. (*Jpn Circ J* 2001; 65: 991–993)

Key Words: Fulminant myocarditis; Immunosuppressive therapy; Intra-aortic balloon pumping; Percutaneous cardiopulmonary support; Polymyositis

Myocardial damage in patients with polymyositis is usually mild, and cardiac involvement has been observed in the chronic stage. We report a patient with polymyositis who suffered fulminant myocarditis. Her cardiac function recovered after immunosuppressive therapy of a large amount of methylprednisolone and intensive therapy, including intra-aortic balloon pumping (IABP) and percutaneous cardiopulmonary support (PCPS).

Case Report

In July 2000, a 54-year-old woman was admitted with dyspnea and a high fever after suffering from proximal limb muscle weakness for 3 weeks. Physical examination showed marked proximal skeletal weakness (III/V), a regular rhythm of 135 beats/min, blood pressure of 100/60 mmHg, and a temperature of 38.2°C. Auscultation revealed widespread, coarse crackles and a systolic blowing murmur (grade III/VI) at the cardiac apex. Chest X-ray revealed a cardiothoracic ratio of 60% and obvious pulmonary edema. The electrocardiogram showed poor R progression in leads V_{2–4}, and mild ST-T depression in V₁ and V₂ (Fig 1). Laboratory studies revealed a white blood cell count of 3,200/mm³ and C-reactive protein of 3.0 mg/dl, and increased amounts of skeletal and cardiac muscle enzymes (creatinine kinase (CK) 2,103 IU/L, aldolase 34.6 U/L, myoglobin 1,710 ng/ml, and CK-MB 206 IU/L). Two-dimensional echocardiography showed hypokinetic wall motion (left ventricular ejection fraction 40%).

Polymyositis with cardiac involvement was suspected, and the patient was placed on prednisolone 20 mg/day and furosemide 20 mg/day. On the third day of hospitalization,

she was transferred to a coronary care unit because the ECG showed frequent ventricular premature contractions with couplets. Although her levels of cardiac muscle enzyme did not increase (CK-MB 26 IU/L), the ejection fraction deteriorated to 20%, and the hemodynamics, as measured by a Swan–Ganz catheter, showed a cardiac index (CI) of 2.3 L·min⁻¹·m⁻², a stroke volume index (SVI) of 19 ml·beat⁻¹·m⁻² and pulmonary capillary wedge pressure (PCWP) of 12 mmHg. Dopamine (5 µg·kg⁻¹·min⁻¹) and dobutamine (5 µg·kg⁻¹·min⁻¹) were given intravenously. However, after 1 h, her blood pressure fell to 60/30 mmHg, CI to 1.4 L·min⁻¹·m⁻², SVI to 10 ml·beat⁻¹·m⁻² and her PCWP was elevated to 22 mmHg. She was diagnosed with

Fig 1. The electrocardiogram on admission is shown. Poor R progression in V_{2–4}, ST depression in V₁ and V₂, and a couplet of ventricular premature beats were shown. No significant changes were observed on the sixth day.

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Fig 2. M-mode echocardiography pretreatment and post-treatment is shown. Left ventricular wall motion recovered following intra-aortic balloon pumping and percutaneous cardiopulmonary support.

Fig 3. Histopathology of the myocardial autopsy showed diffuse mononuclear cell infiltration and myocyte necrosis. The left ventricle showed scattered areas of fibrosis replacing that of cardiac muscle fibers (hematoxylin-eosin (H&E) stain).

Fig 4. Histopathology of the biceps muscle of the arm. Degeneration and regeneration of muscle fibers with inflammatory cell infiltration were observed (H&E).

Fig 5. Histopathology of the myocardial autopsy showed mural thrombus in the left ventricle (H&E).

polymyositis in fulminant myocarditis and intensive therapy, respiratory intubation, IABP and PCPS were instituted.

Immediately after starting PCPS, sustained ventricular tachycardia suddenly occurred. Heparin was used because the anticoagulation time was more than 250 s. Methylprednisolone (2 g/day) was started intravenously instead of oral prednisolone. On the third day after PCPS, the ejection fraction, as measured by echocardiography, recovered to 50% (Fig 2). No significant changes in the ECG were observed during hospitalization (Fig 1). The next day, a massive cerebral embolism occurred, and the patient died subsequently on the 10th day of hospitalization.

An autopsy was permitted by her family, but was limited

only to a section of the heart and the biceps muscle of her arm. After making a minimal incision in the chest wall, the left ventricular anterior wall was excised immediately after death. The cut surface revealed several small spotty lesions of grayish-white tissue in the myocardium. Microscopic examination showed necroses of muscles and diffuse invasion of mononuclear cells, which mainly consisted of lymphocytes in both the myocardium (Fig 3) and the biceps muscle of the arm (Fig 4). Scattered areas of fibrosis replacing that of cardiac muscle fibers were observed (Fig 3). Mural thrombus in the left ventricle was observed (Fig 5). There was no predominant rise in the virus antibody titers between the first and tenth day of hospitalization. The clinical course is presented in Fig 6.

Discussion

Although the incidence of cardiac involvement in polymyositis is 25–70%, it is usually asymptomatic and associated with a mild clinical course.^{1,2} The basic abnormality is an inflammatory process with necrosis and fibrosis of the myocardium, which is similar to the pathological changes seen in the skeletal muscle.³ Therefore, in past reports, cardiac involvement has usually been observed in the chronic stage of polymyositis. Steroids are used mainly to treat polymyositis and also for cardiac involvement.

Previous reports have indicated only 1 case of polymyositis presenting with fatal acute myocarditis as the principal manifestation, and involved fatal arrhythmia 24 h after treatment was initiated using a corticosteroid.⁴ In the present case, fulminant myocarditis was diagnosed on the basis of clinical features, including the presence of severe hemodynamic compromise, and the rapid onset of symp-

Fig 6. Clinical course of the patient. On the third day, the patient was transferred to a coronary care unit. Her cardiac function and hemodynamics rapidly improved after a large dose of methylprednisolone was given as pulse treatment. A massive cerebral embolism occurred on the 6th day and she died on the 10th day of hospitalization.

toms and fever.⁵ Polymyositis was diagnosed based on meeting the criteria of 3 items from a possible 5: muscle weakness, elevated levels of muscle enzyme, a diagnostic muscle biopsy, a characteristic skin rash, and a typical electromyogram. In the present study, polymyositis was diagnosed according to the findings of muscle weakness, elevated levels of muscle enzyme, and biopsy. A simultaneous second process causing acute idiopathic myocarditis in the patient could not be excluded completely. However, the chronological course, mononuclear cell infiltration, which mainly consisted of lymphocytes for both cardiac and skeletal muscle, and the effectiveness of immunosuppressive therapy suggested that the myocarditis was associated with polymyositis. It has been reported that the long-term outcome for fulminant myocarditis is better than for acute non-fulminant myocarditis and if residual cardiac fibrosis is mild.⁶ Therefore, it is very important to support acute phase myocarditis with mechanical assistance such as IABP and PCPS when it is difficult to control using other conventional therapies.⁷⁻⁹ The mechanism of rapid and reversible cardiac dysfunction is not clear, but substances related to inflammation, such as nitric oxide, cytokine and autoantibody, are thought to be involved.¹⁰ Mason et al have reported that immunosuppressive therapy, such as steroids, is not effective for fulminant myocarditis.¹¹ In the present case, however, the patient's cardiac function rapidly improved after a large dose of methylprednisolone had been administered as pulse treatment. However, as cases of polymyositis complicated by fulminant myocarditis are seldom reported, further investigation is necessary to determine whether steroids are an effective treatment.

Incidentally, a left ventricular mural thrombus was formed despite the infusion of heparin. As a possible mechanism, we believe that blood stasis resulting from the left

ventricular wall motion abnormality and hypercoagulability because of inflammation may have played an important role in the thrombogenesis. It was likely that recovery of left ventricular wall motion released a mural thrombus resulting in massive cerebral embolism.

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