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 V2–4, 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 (creatine kinase (CK) 2103 IU/L, aldolase 34.6 IU/L, myoglobin 1710 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...
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. 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. 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. 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-
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的症状和发热。多发肌炎被诊断基于符合3项中的5项：肌肉无力、肌肉酶水平升高、肌肉活检结果、特征性皮肤疹、以及典型的肌电图。在本研究中，多发肌炎被诊断基于肌肉无力、肌肉酶水平升高，以及活检结果。同时存在引起急性自身免疫性心肌炎的第二过程未能完全排除。然而，根据临床过程，单核细胞浸润，主要由淋巴细胞构成，以及免疫抑制治疗的有效性表明，心肌炎可能与多发肌炎有关。已经报告，重症心肌炎的长期预后优于非重症心肌炎，如果残留的心脏纤维化轻微。因此，对于重症心肌炎，急性期心肌炎应支持机械协助，如IABP和PCPS，当难以通过其他常规治疗控制时。机制仍不清楚，但与炎症相关物质，如一氧化氮、细胞因子和自身抗体可能参与。


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.

References