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A survival case of invasive thymoma accompanied by acute fulminant myocarditis

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Abstract

Thymomas are associated with a wide spectrum of autoimmune paraneoplastic diseases. Here we report the case of 31-year-old male with invasive thymoma, myasthenia gravis, polymyositis, and acute fulminant myocarditis that presented with cardiogenic shock requiring intra-aortic balloon pumping and percutaneous cardiopulmonary support. Corticosteroid therapy was effective. To our knowledge, this is the first case of thymoma with acute fulminant cardiomyositis that was successfully treated by assisted circulation and corticosteroids, despite a poor prognosis.

Introduction

There are numerous reports on the association of the thymomas with a wide spectrum of autoimmune paraneoplastic diseases [1], however, myocarditis accompanied with thymoma is rare [2–5]. Here we report a patient with invasive thymoma with myasthenia gravis, polymyositis and acute fulminant myocarditis that manifested as cardiogenic shock that required intra-aortic balloon pumping and percutaneous cardiopulmonary support. Corticosteroid therapy was successfully administered. This is the first case, to our knowledge, survival from thymoma with acute fulminant myocarditis. We have discussed treatment strategies for this lethal disease combination.

Case Report

A 31 year-old male with a history of thymoma (type B2, stage IVa) diagnosed at 28 year-old, myasthenia gravis and polymyositis, had been treated with chemotherapy (four amurubicin courses, three cisplatin plus VP-16 courses, and three ADOC, including cisplatin, doxorubicin, vincristine, and cyclophosphamide) and tumour debulking. He was also on long-term prednisolone for myasthenia gravis and polymyositis. Prednisolone dose tapering to 25 mg resulted in a relapse of polymyositis, necessitating re-hospitalization. On the fourth day of hospitalization, he developed ventricular tachycardia appeared with cardiogenic shock requiring intra-aortic balloon pumping and percutaneous cardiopulmonary support. Echocardiography revealed diffuse hypokinesis, with decreased in ejection fraction to approximately 20%. Laboratory tests revealed an elevated creatinine phosphokinase level 6300 U/L and a creatinine kinase MB level of 233 U/L. Chest computed tomography demonstrated known left pleural disseminated lesions of thymoma enlarged, however, no myocardial invasion. Cardiac catheterization and biopsy revealed no coronary artery stenosis, while cardiac biopsy revealed lymphocytic myocarditis (Fig. 1). Infection and drugs were excluded as viral titers were negative and the total doxoru-

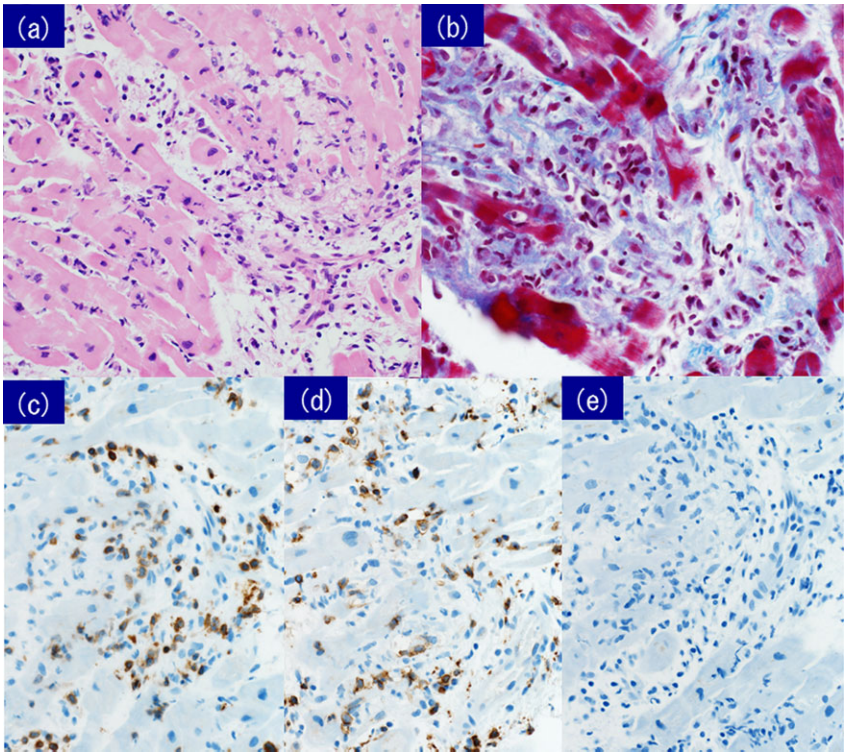

Figure 1. Endomyocardial biopsy specimens showing diffuse interstitial lymphocytic infiltration with myocyte damage (a: Hematoxylin-Eosin staining, b: Masson trichrome staining). Immunohistochemical staining is positive for CD3 (c) and CD8 (d) which are T cell marker, but negative for CD20 (e) which is B cell marker. These findings indicate the presence of T cell-mediated myocarditis.

bicin dose was less than 350 mg/m^2. These findings were consistent with myocarditis as a paraneoplastic disease. Steroid pulse therapy (methylprednisolone, 1 g/day for three days) was administered, followed by prednisolone (1 mg/kg/day). Intra-aortic balloon pumping and percutaneous cardiopulmonary support were terminated four days after the initiation of steroid pulse therapy, with a recovery of ejection fraction to 74%. The acute myocarditis stabilized and irradiation therapy for residual pleural dissemination was initiated. Thymoma responded well to the irradiation and prednisolone therapy, and there were no sign of relapse, up to ten months following this episode of cardiogenic shock due to acute fulminant cardiomyositis.

Discussion

Twenty-two patients of thymoma-associated paraneoplastic myocarditis have been reported so far [2–5]. All patients exhibited WHO type B1 and B2 thymoma and myasthenia gravis. At onset, three patients were on tapering steroid therapy, five had relapsed thymoma, and two had been triggered by chemotherapy [3]. Histopathology revealed giant cell myocarditis in 20 patients and lymphocytic myocarditis in two. These findings were mainly obtained from postmortem analysis as almost all had died suddenly or during the acute phase. The treatment possibilities of this disease have therefore not been well described in the literature. It is suggested that an autoimmune process involving activated T lymphocytes may be important in myocarditis development [2, 5]. Therefore, steroids, immunosuppressants, and immunoglobulin have been used for cardiomyositis [2, 4], although their effectiveness has not been established. Our patient survived because he was hospitalized early; received timely antiarrhythmic drugs, defibrillation, and percutaneous cardiopulmonary support; and his condition responded well to corticosteroids. In conclusion, thymoma with acute fulminant myocarditis is a clinically challenging condition, although corticosteroid therapy and assisted circulation appear to be effective in its treatment.

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Disclosure Statements

No conflict of interest declared.

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

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