CASE REPORT

Fatal giant cell myocarditis after resection of thymoma

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Abstract

A case of fulminant, fatal myocarditis occurring 10 days after resection of a benign medullary thymoma is described. A rare association between thymoma and giant cell myocarditis is recognised, but fulminant presentation so soon after removal of thymoma has not previously been reported.

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Case report

A 57 year old man who had no previous history of cardiac disease presented with recurrent chest infections and was found to have a mass with chest radiography. Computed axial tomography showed a well circumscribed mass located in the left hemithorax (fig 1). An electrocardiogram, echocardiogram, and routine blood tests (including white cell count and erythrocyte sedimentation rate) were normal. A biopsy specimen of the mass showed

the presence of a thymoma. Surgical resection was performed at the Department of Cardiothoracic Surgery, Royal Brompton Hospital. A large (1 kg, 17 × 12 cm), well circumscribed tumour filling the left hemithorax was removed. This was adherent to, but did not invade, the anterior pericardium (absence of pericardial infiltration by tumour was confirmed histologically). A disc of pericardium was resected with the tumour, and radical mediastinal clearance performed. Histological analysis showed a mixture of mature lymphocytes and epithelial cells showing no atypia or mitotic activity. The tumour epithelial cells had the appearance of medullary spindle cells which was consistent with a benign thymoma. The patient made an uneventful recovery and was discharged.

Ten days after surgery he was transferred urgently to the Royal Brompton Hospital with a 24 h history of breathlessness at rest. He denied chest pain. On examination he had a regular tachycardia of 120 beats/min, and the jugular venous pressure was elevated to 5 cm. The blood pressure was 130/70 mm Hg, heart sounds were normal, and no murmurs or added sounds were heard. An electrocardiogram (fig 2) showed sinus tachycardia with widespread ST segment elevation. An echocardiogram showed dilatation of the left and right ventricular cavities, and globally impaired ventricular contraction. No pericardial fluid or valvular abnormality was detected. Serum potassium, calcium and magnesium levels were normal. Ventricular tachycardia (180 beats/min) supervened shortly afterwards. DC cardioversion under general anaesthesia produced no sustained return to sinus rhythm. An intravenous bolus of lignocaine was also unsuccessful, and therefore a central venous infusion of amiodarone was started. Ventricular tachycardia persisted and the patient developed cardiogenic shock despite inotropic support in the form of dobutamine and adrenaline. Because of the rapid deterioration in the absence of a diagnosis, and to rule out the possibility of massive pulmonary embolus, emergency thoracotomy was performed on the ward. Intracardiac pressures were normal, and there was no evidence of pulmonary embolus or myocardial infarction. Ventricular tachycardia persisted in spite of attempts at overdrive pacing. The patient was

Figure 1 Computed axial tomogram showing a large, well circumscribed mass located in the anterior left hemithorax.

Figure 2 Electrocardiogram showing sinus tachycardia with widespread ST segment elevation.

Figure 3 Myocardial histological specimen obtained at postmortem examination showing giant cell myocarditis. Myocytes vary in size and appear degenerate. There is an infiltrate of lymphocytes, plasma cells, epithelioid cells, and giant cells (arrow) between and around the myocytes. Haematoxylin and eosin, originally $\times 350$.

transferred to the intensive care unit where, despite aortic balloon counterpulsation, ventilation, and continued inotropic support, he died 24 h later.

The heart weighed 447 g at postmortem examination. The myocardium was noted to be uniformly soft and pale. No abnormality of the coronary circulation or heart valves was present. Other organs were essentially normal and there was no evidence of recurrent or metastatic thymoma. Histological examination of the myocardium showed diffuse myocardial necrosis. Plasma cells, lymphocytes, epithelioid cells, and giant cells were seen to infiltrate around the necrotic myocytes (fig 3). These appearances are characteristic of giant cell myocarditis.

Discussion

Giant cell myocarditis generally affects young to middle aged adults of either sex. It is char-

acterised by acute heart failure, ventricular arrhythmias, or heart block which progress rapidly to death usually within 10 days.¹ More widespread use of endomyocardial biopsy has allowed diagnosis of milder cases, and long-term survival has been reported.² Giant cell myocarditis usually occurs in isolation, but has been reported in association with a variety of autoimmune diseases^{1,3} and chronic infections.⁴⁻⁶ Although a giant cell infiltrate is a prominent feature, it is histologically distinct from Wegener's granulomatosis or sarcoidosis. It is diagnosed in approximately 1% of patients with thymoma.¹

The aetiology of giant cell myocarditis is unknown. Infectious causes have been sought but not consistently found.¹ There is some evidence to suggest an autoimmune process⁷⁻⁹ and activated T lymphocytes seem to be important for myocarditis to develop.⁸ Myocarditis has been reported to recur in transplanted hearts.^{10,11}

Giant cell myocarditis occurring so soon after resection of thymoma has not previously been reported. There was no evidence of active myocarditis before surgery in this patient. It is tempting to speculate that removal of the thymoma may in some way have triggered myocarditis. Theoretically this could be caused by removal of the inhibitory influence of suppressor T lymphocytes, permitting an aggressive inflammatory process. Anecdotal reports and animal studies suggest that immunosuppressive treatment may be effective,^{7,11,12} but there are no randomized controlled trials to support this. Although it is unlikely that aggressive immunosuppression would have altered the outcome in this case, increased awareness of giant cell myocarditis in association with thymoma may lead to early recognition and treatment.

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