Sudden Cardiac Death from Hypertrophic Cardiomyopathy and Acute Idiopathic (Fiedler’s) Myocarditis: Autopsy Report

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A 45-year-old man with a rapidly deteriorating heart condition died suddenly, two hours after admission, after resistance to attempts of cardiac pacing. At autopsy, the heart weighed 600g with asymmetric septal hypertrophy. Histological examination revealed an extensive and diffuse disarray of myocardial fibers in the ventricular septum and in the free wall of both ventricles. Pronounced mononuclear cell infiltrations and interstitial edema were distributed widely in both ventricles. There were few abnormal findings in the other organs. The diagnosis was hypertrophic cardiomyopathy accompanied by Fiedler’s myocarditis. Such a case appears to be rare.

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Key words: Fiedler’s myocarditis, asymmetric septal hypertrophy

Introduction

Fiedler’s myocarditis, first described by Fiedler in 1899 (1), is characterized clinically by a sudden onset and a rapidly fatal outcome, and pathologically inflammatory lesions in the interstitial tissue of myocardium. Hypertrophic cardiomyopathy is also one of the common cause of sudden death (2, 3). Our patients died suddenly after resisting cardiac pacing and other intensive treatment for two hours after admission. Autopsy of the heart showed asymmetric septal hypertrophy and diffuse disarray of myocardial fibers. Pronounced mononuclear cell infiltration was observed in the interstitial tissue of myocardium. Our diagnosis was hypertrophic cardiomyopathy accompanied by Fiedler’s myocarditis. Although case reports of hypertrophic cardiomyopathy accompanied by coronary heart disease are numerous, those showing an accompanying acute myocarditis are rare. There are no previous autopsy reports of hypertrophic cardiomyopathy accompanied by Fiedler’s myocarditis. In this report, we investigated the possible cause of sudden death in hypertrophic cardiomyopathy.

Case Report

A 45-year-old man was admitted in a state of disturbed consciousness. The patient had fever and epigastralgia on the evening prior to admission, which did not resolve overnight. Around noon on the next day, he suffered intrathoracic pain. He then became comatose. In the ambulance, he regained consciousness transiently, but was soon comatose again. There was no past history of hypertension or heart disease in the patient or his family. The patient’s condition for several days before the onset was unknown, and he had not had an electrocardiogram (ECG) during previous the ten years.

On admission, the patient was comatose (Japanese coma scale II-2) and had a temperature of 36.2°C, pulse 35/min and irregular, blood pressure 90 mmHg on palpitation, no jaundice or anemia of conjunctiva, and no distension of the jugular vein. Heart sounds were diminished but no heart murmurs or extra sounds were noted. Rales were not heard in the bilateral lung fields. The liver and spleen were not palpable. There was no peripheral edema or skin eruptions.

Laboratory data on admission are listed in Table 1. White blood cell count (WBC), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were evaluated. Biochemical tests indicated increases in transaminase (GOT, GPT), lactate dehydrogenase (LDH), creatine kinase (CK), blood urea nitrogen (BUN), serum creatinine (SCr), serum amylase (SAmyl), blood sugar (BS) and potassium. Arterial blood gas analysis showed marked metabolic acidosis. ECG revealed bradycardia with inverted T waves in II, III, and aVF, and deep inverted T waves accompanied by ST segment depression in V2–V6. The frequent appearance of a polymorphic ventricular extrasystole resulted in chaotic rhythm (Fig. 1). These findings indicated ischemic and/or hypertrophy of the myocardium and also
Table 1. Laboratory Data on Admission

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Test</th>
<th>Result</th>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC</td>
<td>37,800/mm³</td>
<td>BS</td>
<td>247 mg/dl</td>
<td>pH</td>
<td>6.678</td>
</tr>
<tr>
<td>RBC</td>
<td>4.57x10⁶/mm³</td>
<td>BUN</td>
<td>53 mg/dl</td>
<td>PCO₂</td>
<td>21.5 mmHg</td>
</tr>
<tr>
<td>Hb</td>
<td>12.5 g/dl</td>
<td>SCr</td>
<td>5.0 mg/dl</td>
<td>PO₂</td>
<td>75.4 mmHg</td>
</tr>
<tr>
<td>Ht</td>
<td>39.9%</td>
<td>CK</td>
<td>386 IU/l</td>
<td>HCO₃⁻</td>
<td>-2.6 mmol/l</td>
</tr>
<tr>
<td>Plt</td>
<td>48.0x10⁴/mm³</td>
<td>MB</td>
<td>90 IU/l</td>
<td>BE</td>
<td>-35.5</td>
</tr>
<tr>
<td>TP</td>
<td>6.5 g/dl</td>
<td>MM</td>
<td>296 IU/l</td>
<td>PCO₂</td>
<td>21.5 mmHg</td>
</tr>
<tr>
<td>GOT</td>
<td>901 IU/l</td>
<td>Na</td>
<td>136 mEq/l</td>
<td>O₂ SAT</td>
<td>78.0%</td>
</tr>
<tr>
<td>GPT</td>
<td>77 IU/l</td>
<td>K</td>
<td>5.8 mEq/l</td>
<td>(room air)</td>
<td></td>
</tr>
<tr>
<td>LDH</td>
<td>1,121 IU/l</td>
<td>Cl</td>
<td>92 mEq/l</td>
<td></td>
<td></td>
</tr>
<tr>
<td>T-Bil</td>
<td>0.7 mg/dl</td>
<td>ESR</td>
<td>72 mm/h</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SAmyl</td>
<td>276 IU/l</td>
<td>CRP</td>
<td>3+</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

failure of other organ systems due to cardiogenic shock. Chest X-rays showed no congestion in the lung fields (cardiothoracic ratio: 53%). While recording the ECG at the outpatient clinic, the patient suddenly stopped breathing and went into cardiac arrest. Emergency resuscitation was performed, and he was admitted and placed on an artificial respirator. Sinus arrest and drug-resistant ventricular tachycardia appeared frequently, and blood pressure therefore remained at a low level. Under these conditions, temporary intravenous pacing and intraaortic balloon pumping were performed. A pacing rhythm of 70/min was maintained for a few minutes. However, the pacing threshold increased gradually, and the patient became resistant to cardiac pacing. Chaotic rhythm then ensued and he died.

At autopsy, the heart weighed 600 g. Concentric hypertrophy was observed, along with asymmetric septal hypertrophy (ventricular septal: 24 mm, posterior wall: 18 mm; septal/posterior wall = 1.33) but there was no left ventricular dilatation (Fig. 2). The subaortic septal band was not observed at the upper part of the septum. Postmortem angiography revealed no occlusion or stenosis of the coronary vasculature. Gross examination revealed no infarction foci or change in the valves. Pericardial effusion was also absent. Histological examination revealed hypertrophic myocardial fibers with disarray in the middle layer of the ventricular septum and also in the free wall of both ventricles (Fig. 3, Fig. 6a). Mild intimal hypertrophy of small arteries was noted in the septal myocardium. A pronounced infiltration of mononuclear cells, and a small number of monocytes.}

![ECG on admission.](image)

**Fig. 1.** ECG on admission. Note bradycardia with inverted T waves in II, III, aVF and deep inverted T waves accompanied by ST segment depression in V2–V6. QRS duration of 0.14 second, QTc of 0.52 second, SV1 + RV5 = 42 mm.
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**Fig. 2.** Macroscopic findings observed in the short axis section of the heart. Note concentric hypertrophy and asymmetric thickening of the septum.

**Fig. 3.** Microscopic findings obtained from the middle layer of the ventricular septum. Disarray of the fibers is also observed in the free wall of both ventricles (hematoxylin phosphotungstate, ×50).

**Fig. 4.** Microscopic findings obtained from the left ventricular free wall. Note severe inflammatory cell infiltration and edema in the myocardial interstitial tissue (HE stain, ×50).

**Fig. 5.** Note the infiltration of lymphocytes mainly by T cells (immunohistochemical staining using a paraffin specimen, ×100).

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eosinophils and mast cells, with interstitial edema, was observed mainly in the middle layer of the left ventricle and ventricular septum (Fig. 4, Fig. 6b). Examination using monoclonal antibodies disclosed that most lymphocytes were T cells (MT-1 positive >> MB-1 positive) (Fig. 5). Slight degeneration and necrosis of myocytes were observed. Masson’s Trichrome staining revealed edema around the blood vessels, but fibrous changes were minimal. There were no findings of necrotic angiitis, granuloma, or giant cell infiltration. Inflammation in the endocardium or epicardium was present in a very limited area (Fig. 6b). There was mild congestion in the spleen, kidney and small intestine. The lungs weighed 250 g (left) and 380 g (right). There was a small hemorrhagic infarction in the right lobe. Moderate fatty degeneration of the liver was observed. There was no evidence of inflammatory changes except for the interstitial tissue of the myocardium. Mild arteriosclerosis, compatible with the patients age was noted in the aorta and coronary arteries. Histological evaluation did not indicate hypertensive changes. The present case was diagnosed clinicopathologically as hypertrophic cardiomyopathy accompanied by Fiedler’s myocarditis.

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**Discussion**

In 1899, Fiedler reported four cases of fulminant myocarditis with sudden onset and rapidly fatal outcome (1). Histological features primarily include inflammatory changes occurring in the myocardial interstitial tissue which spare the endocardium,
question of whether inflammation has occurred by chance in a hypertrophic ventricular septum or whether inflammation itself has caused the asymmetric septal hypertrophy. There is a case report of fulminant myocarditis was caused by asymmetric septal hypertrophy due to marked interstitial edema and inflammatory cell infiltration in the ventricular septum (12). Reversible asymmetric septal thickening was also demonstrated by echocardiogram in a case of acute myocarditis (13). According to Okada, disarray of myocardial fibers could occur after fibrous changes due to myocarditis (14). However, the autopsy findings of the present case revealed extensive infiltration of acute inflammatory cells and diffuse disarray of myocardial fibers in both the ventricular septum and ventricular free wall. The left ventricular wall thickening did not correlate well with the intensity of the inflammatory changes.

Because the patient was comatose and had severe inflammation, metabolic acidosis, and abnormal biochemical data at admission, we considered as possible causes, multiorgan failure due to cardiogenic shock, drug-induced or septic shock, and viral inflammation involving multiple organs. At autopsy, we observed no severe eosinophilic infiltrate that would suggest hypersensitivity myocarditis. There were no apparent inflammatory changes in the other organs or chronic dysfunction in the liver or kidney.

Generally, fulminant myocarditis causes cardiogenic shock, associated with severe myocardial necrosis and impaired wall motion. However, the present case had no severe myocardial necrosis or pulmonary congestion on histologic observation, suggesting a very rapid clinical course. We speculated that the direct cause of sudden death had been acute pump failure due to a chaotic ventricular rhythm. Although there have been numerous reports of rescue from Adam-Stokes attacks caused by acute myocarditis by applying cardiac pacing (15), fulminant myocarditis has often been reported to be unresponsive to such pacing (16, 17). We considered the possible mechanism of resistance to the cardiac pacing and the subsequent cardiac arrest. It may be that the pacing threshold increased because of infiltration of inflammatory cells and interstitial edema due to myocarditis, or because of failure of the electrolytic environment in cardiac cells due to severe acidosis.

We concluded that asymmetric septal hypertrophy was due to hypertrophic cardiomyopathy, and diagnosed hypertrophic cardiomyopathy accompanied by Fiedler's myocarditis. To our knowledge, there has been one report of hypertrophic cardiomyopathy accompanied by acute benign myocarditis (18), but there is no autopsy report of hypertrophic cardiomyopathy accompanied by Fiedler's myocarditis; this appears to be the first such report.

References

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