

A Case of Left Ventricular Endomyocardial Fibrosis

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The patient was a 29-year-old woman. When she consulted a local physician with chief complaints of fever and fatigue of the extremities, cerebral infarction was detected on MRI, in addition to abnormalities found on ECG. Ultrasonic cardiography revealed the presence of a tumor in the left ventricle. Therefore, tumorectomy and endocardectomy were performed under extracorporeal circulation based on a diagnosis of cardiac tumor. Inflammatory cell infiltration into the ventricular wall was pathologically confirmed, and eosinophilia was observed preoperatively. Therefore, the patient was diagnosed as having endomyocardial fibrosis, which is rarely observed in Japan. The postoperative course of this patient was satisfactory, and the eosinophil count was normalized postoperatively. At present, this patient is being followed at the outpatient clinic. (Ann Thorac Cardiovasc Surg 2002; 8: 173–6)

Key words: endomyocardial fibrosis, endocardectomy

Introduction

Endomyocardial fibrosis (EMF) is a progressive restrictive cardiomyopathy frequently observed in tropical and subtropical regions. However, EMF is rarely observed in Japan.¹⁾ EMF is characterized by fibrous endomyocardial hypertrophy and calcification, and mural thrombosis in the bilateral or hemilateral ventricle. Since EMF patients develop heart failure during the advanced stage of the disease, the mean survival period was reported to be 2–4 years after the manifestation of heart failure symptoms.²⁾ Currently, there is no effective medical treatment for EMF, and symptomatic therapy is mainly used to treat EMF. However, some previous studies reported that surgical treatment was useful for treating EMF.^{3,4)} In the present study, we performed endocardectomy in a patient with EMF, and favorable results were obtained. Here, we report the course of this patient, together with some bibliographical comments.

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Case Report

The patient was a 29-year-old woman with chief complaints of fever and fatigue of the extremities. Since fever and fatigue of the extremities were noted at the beginning of October 1999, the patient consulted a local physician. She also reported hearing myodesopsia. Cranial MRI was performed, and cerebral infarction was detected. ECG abnormalities were also detected, and the patient was referred to the cardiovascular department of our hospital. Since ultrasonic cardiography demonstrated a tumor in the left ventricle, the patient was referred to our department. The patient developed atopic dermatitis at the age of 20 and Basedow's disease was diagnosed at the age of 25, which is still being treated. Family history was not contributory. At admission, the patient was 154.5 cm tall, and weighed 49.6 kg. The pulse rate was 98/min and regular. The blood pressure level was 110/52 mmHg. There was no cardiac murmur or rale heard. There were no findings of anemia, jaundice, hepatosplenomegaly, or edema. Her blood level readings were white cell count of 8,400/mm³, neutrophil ratio of 22% (1,848/mm³), LDH levels of 332 U/l, CRP levels of 0.19 mg/dl, and eosinophilia were observed. A chest X-ray showed no cardiomegaly with a cardiothoracic ratio was 45%, and no

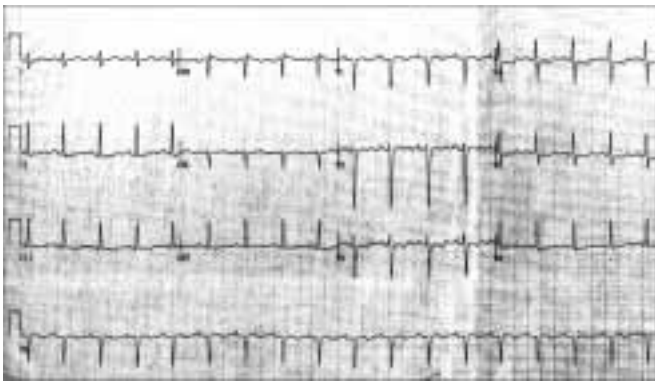


Fig. 1. Low electric potentials and slight ST depressions were observed in chest and V₄₋₆ leads on ECG.

abnormal findings such as enhanced pulmonary vascular shadows, pleural effusion collection, or calcification were observed. ECG findings were low electric potentials and slight ST depressions were observed in chest and V₄₋₆ leads (Fig. 1). According to echocardiography, there were no findings of diastolic disorder and EF was 63%. However, a tumorous lesion with internal nonhomogeneity measuring 34×34 mm was observed in the apical region (Fig. 2a, b). According to cardiac catheterization, abnormal findings were not observed in the bilateral coronary arteries. In addition, neither tumor stain nor nutrient vessels were detected. Therefore, surgery was performed on December 10, 1999 based on a diagnosis of cardiac tumor.

Intraoperative findings

After median sternotomy, extracorporeal circulation was initiated by pumping blood from the superior and inferior vena cava and returning it through the ascending aorta. Under cardiac arrest, the left ventricle was resected parallel to the left anterior descending branch of the coronary artery. The resected myocardium was degenerated and the color had changed to yellowish brown. The tumor protruded toward the ventricular lumen (Fig. 3). The interior of the tumor was fragile, and caseous degeneration was observed. Therefore, the tumor and the tissue around the thick endomyocardium were completely resected. We could confirm normal myocardium tissue in the surgical margin before closing the left ventricle. After surgery, the patient was easily weaned from extracorporeal circulation. During 5 hours of surgery, the aorta was blocked for 72 min, and the patient was supported by extracorporeal circulation for 133 min. Postoperative pathological findings, part of the resected tissue speci-

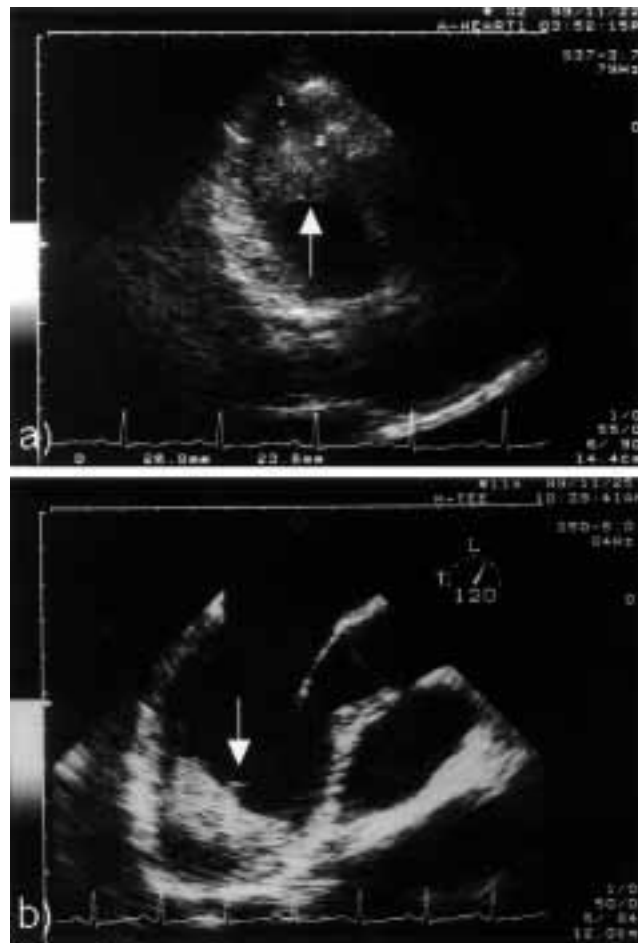


Fig. 2. EF was 63%, and there were no findings of diastolic disorder. However, a tumor with internal nonhomogeneity measuring 34×34 mm was observed in the apical region (↑, ↓). (a: transthoracic approach, b: transesophageal approach)

mens included organizing thrombi, in which eosinophilic granules were observed. In addition, organization was also observed in the endocardium to which the thrombi adhered. Inflammatory changes in the resected myocardium suggested the presence of endomyocarditis accompanying mural thrombosis (Fig. 4a, b). The patient's postoperative course was uneventful. The eosinophil count was normalized postoperatively, and the patient was discharged from the hospital 19 days after surgery. Currently, the patient has returned to her daily life without any signs of recurrent EMF.

Discussion

EMF is a progressive restrictive cardiomyopathy frequently observed in tropical and subtropical regions, and is rarely observed in Japan.¹⁾ EMF occurs in the bilateral

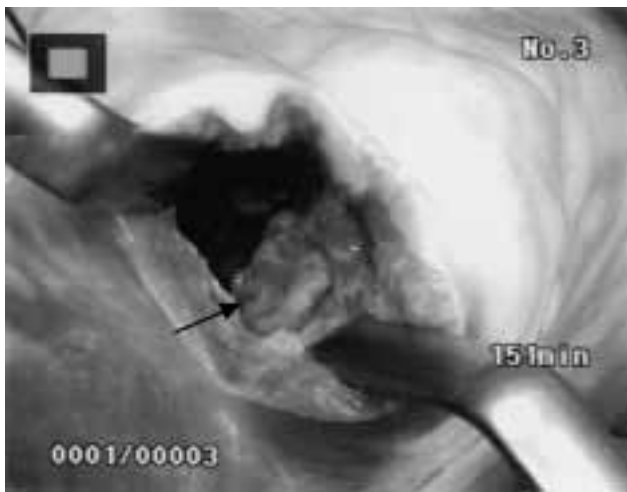


Fig. 3. The resected myocardium was degenerated and the color had changed to yellowish brown. The tumor protruded toward the ventricular lumen. In addition, the interior of the tumor was fragile, and caseous degeneration was observed (→).

or hemilateral ventricle, and is characterized by fibrous atrophy of the apical wall, subvalvular tissues such as the papillary muscle, endocardium, and subendocardial myocardium, resulting in valve regurgitation and ventricular diastolic disorder. Since EMF patients develop intractable heart failure during the advanced stage of the disease, the mean survival period is reported to be approximately 2-4 years after the manifestation of heart failure symptoms.²⁾ It has been speculated that EMF is caused by serotonin

contained in bananas harvested in the tropical regions,⁵⁾ or by eosinophils and immunological abnormalities.⁶⁾ However, the etiology of EMF remains unclear. Most EMF patients were reported to complain of heart failure symptoms, beginning with fever, physical weakness, and fatigue.⁷⁾ However, careful examinations are required because there is no characteristic symptom of EMF. Previously reported cases of EMF were diagnosed based on the following findings: cardiac dilatation and apical calcification on chest X-ray; and electrocardiographic findings of low electric potentials, flat T waves, atrial fibrillation, atrioventricular block, and ventricular tachycardia.⁸⁾ Echocardiography showed findings of intraventricular thrombosis, pericardial effusion collection, and increased echo from the epicardium.⁹⁾ In addition, cardiac catheterization showed a dip and plateau of diastolic ventricular pressure and decreased cardiac index. Although some previous studies reported the results of myocardial biopsy from EMF patients, thorough investigation is required to establish a definitive diagnosis because myocardial biopsy may produce false-positive results due to the presence of an embolism. A therapeutic strategy for EMF has not yet been established, and steroid hormone therapy or immunosuppressive therapy is currently being attempted. Since most cases of EMF are therapy resistant, symptomatic therapy is mainly used to treat heart failure and to prevent thrombus formation. However, the prognosis is poor in EMF patients after the manifestation of heart failure symptoms.²⁾ Although some previously

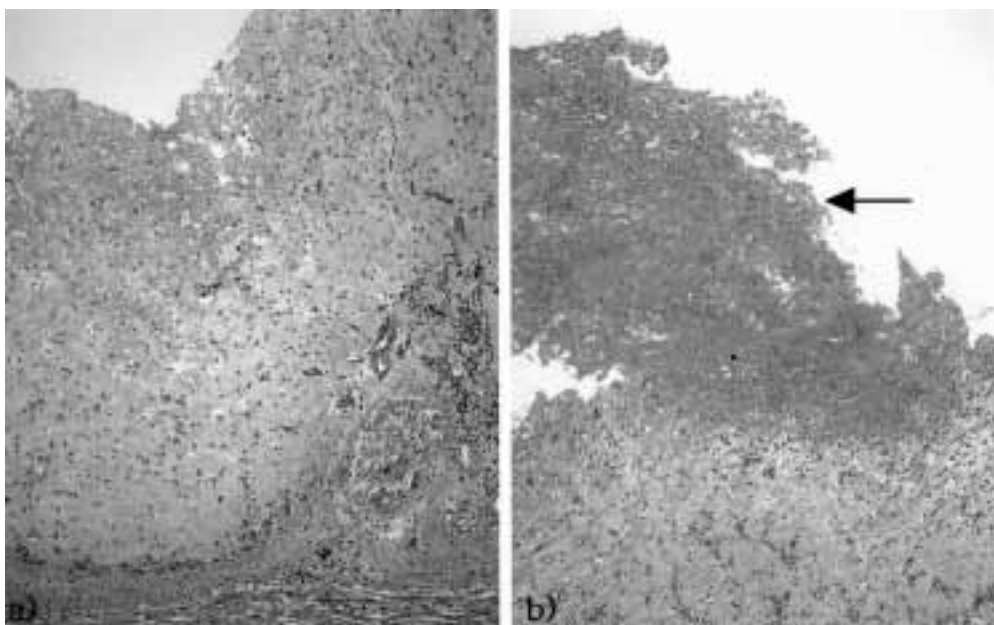


Fig. 4. a: Findings of endomyocarditis were observed, and organizing thrombi were detected in the left ventricular lumen.
b: Fresh thrombus formation was observed in part of the left ventricle.

reported cases of EMF were successfully treated by surgery consisting of endocardectomy and valve replacement, the mortality rate was as high as approximately 20%, probably because most surgically treated cases of EMF were in the advanced stage of the disease.³⁾ However, a better prognosis may be obtained even in EMF patients when tumorectomy can be performed successfully in a relatively limited range of the myocardium. Since early detection of EMF is difficult, only one case of EMF was surgically treated in Japan during the advanced stage of the disease, resulting in difficult postoperative management of the patient.⁴⁾ Although EMF is rarely observed in Japan, it is important to examine patients complaining of agnogenic fever or eosinophilia considering the possibility of EMF in the differential diagnosis.

Moreover, since several cases of recurrent EMF have been reported postoperatively,¹⁰⁾ the postoperative course of EMF patients should be followed carefully.

Conclusions

We encountered a patient with left ventricular endomyocardial fibrosis (EMF), which is rarely observed in Japan. After surgical treatment, favorable results were obtained. Echocardiography may be the most useful examination procedure for diagnosing EMF.

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