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KEYWORDS: cardiomyopathy, idiopathic cardiomyopathy, hypertrophic cardiomyopathy

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AN AUTOPSY CASE OF HYPERTROPHIC CARDIOMYOPATHY SHOWING CLINICAL FEATURES OF DILATED CARDIOMYOPATHY

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Abstract. An autopsy case of hypertrophic cardiomyopathy showing clinical features of dilated cardiomyopathy was reported. The patient was a 60-year-old female complaining of chest discomfort from the age of 40. At autopsy, both ventricles were dilated. Microscopically myocardial loss, fibrosis and disarray of hypertrophic myocardial fibers were observed. The areas showing myocardial disarray were distributed close to the scar-like fibrotic areas. Coronary arteries and intramyocardial arterioles showed minimal stenotic changes.

Key words : cardiomyopathy, idiopathic cardiomyopathy, hypertrophic cardiomyopathy.

Some cases of idiopathic hypertrophic cardiomyopathy (HCM) show clinical features of dilated cardiomyopathy (DCM) in the late stage of the disease (1). In 1979, Maron *et al.* (2) reported some cases of HCM showing chronic congestive heart failure due to extensive myocardial scarring without clinically evident acute myocardial infarction, and ten Cate and Roelandt (3) reported two cases of HCM, which progressed to left ventricular dilatation. In Japan, six autopsy cases of DCM-like HCM have been reported (1, 4, 5), but more cases are required to deduce the distinct clinical and pathological characteristics and etiology of the disease. We reported a case of HCM which showed clinical features of DCM, and discussed it in relation with other reported cases.

MATERIALS AND METHODS

Autopsy was done one hour after the death of the patient. The heart was fixed in 10 % formalin and sliced vertical to the axis. Three whole cut planes of the ventricles and other specimens from both atria, the sinoatrial node and the atrio-ventricular node were processed for paraffin blocks. Sections were stained with hematoxylin-eosin (HE) stain, Masson's trichrome stain and Azan stain. Materials for electron microscopy were fixed in 10 % formalin buffered in 0.1 M phosphate (pH 7.4), refixed in 2 % osmium, dehydrated and embedded in Epon. Thin-sectioned specimens were examined with a JEOL transmission electron microscope (JEM 100S).

CASE

The patient was a 60-year-old female. One of her three daughters, who complained of angina-like chest discomfort on effort, was diagnosed as HCM at the age of 38. Other aspects of the family history were not unusual. The patient was given internal and external radiation for cervical cancer when she was 53 years old. She did not smoke or drink. She had no history of hypertension. Dyspnea on effort, chest discomfort, frequent arrhythmia and coughing at night appeared when she was about 40 years old. She was admitted to Okayama National Hospital at the age of 57 in December, 1981.

Upon admission, a systolic heart murmur at the lower left border of the sternum, dilatation of the jugular veins and bilateral palmar erythema were recognized. Neither the liver nor the spleen was palpable. Blood pressure was about 100/60, and the heart rate was 50-60 per min. The results of blood and urine examinations were almost within normal limits. Chest roentgenography showed cardiomegaly with a 63 % cardiothoracic ratio. ECG showed degree I of atrio-ventricular block, intraventricular conduction delay and premature ventricular contractions. UCG revealed hypokinesis of the left ventricular wall and marked dilatation of the left ventricular cavity. Atypical septal hypertrophy was not demonstrated.

Her condition was tentatively diagnosed as DCM and was controlled by medication with digitalis, diuretics and vasodilators such as α -blockers. In November 1982, she complained of severe abdominal pain. At this time, hepatomegaly of three finger breadths and marked dilatation of the jugular veins were observed. Chest roentgenography revealed a 67 % cardiothoracic ratio, pulmonary edema and congestion. Abdominal pain gradually decreased and disappeared with improvement of the heart failure. In November 1983, cervical cancer relapsed, and external and internal radiation was done. In March 1984, postradiation proctitis appeared, and formation of an artificial anus was performed. In May 1984, fever appeared, and many kinds of antibiotics were administered. Gradually, heart failure with atrial fibrillation and dyspnea became more severe. She expired on July 25, 1984.

At autopsy, the heart weighed 450 g and showed cardiomegaly (Fig. 1). On the cut surfaces of the ventricles, both ventricles were dilated (Fig. 2). The thickness of the ventricular septum was 0.9 cm in the anterior part and 1.3 cm in the posterior part. Small whitish fibrotic foci were scattered in the ventricular septum, and a large fibrotic area existed in the posterior part, which extended from the epicardial aspect. The anterior wall of the left ventricle, the myocardium of which was 0.2 cm in thickness, showed myocardial disappearance and fibrosis from the epicardial aspect. The lateral and posterior wall of the left ventricle, the myocardium of which was 0.8 cm and 1.0 cm in thickness, respectively, was scattered with small whitish fibrotic foci. The right ventricular wall, the myocardium of which was 0.3 cm in thickness, also showed scattering of small fibrotic foci. The papillary

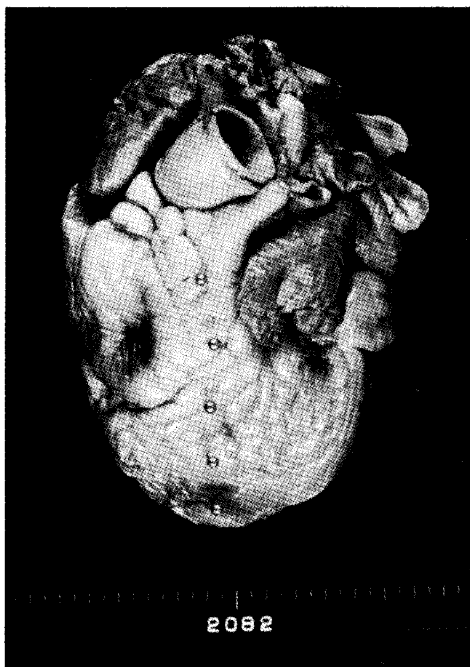


Fig. 1. Anterior view of the heart.

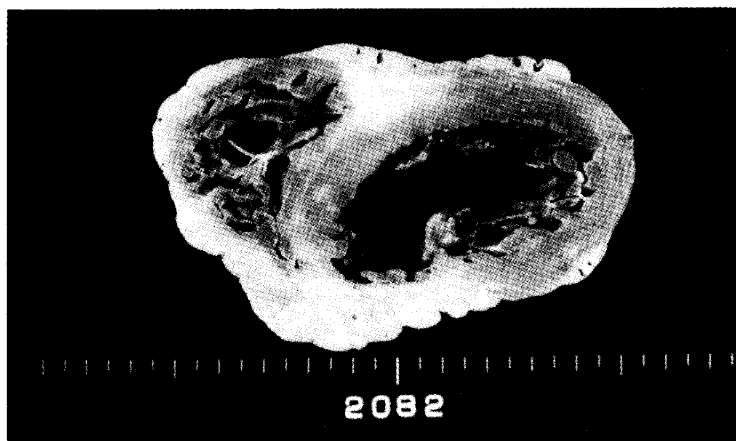


Fig. 2. A cut surface of the heart.

muscles of both ventricles were well developed. Aortic valves, pulmonary valves, mitral valves and tricuspid valves were not remarkable. Both atria and auricula were dilated. The right atrium had a small mural thrombus.

Histologically, the wall of both ventricles and ventricular septum showed fibrosis as follows: (1) Massive disappearance of myocardial fibers and scar-like

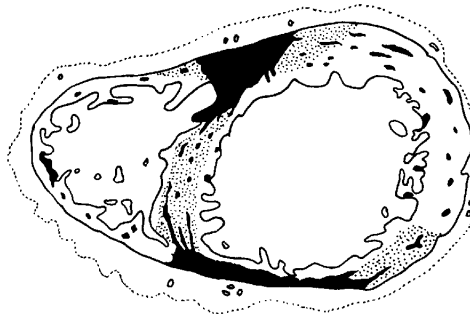


Fig. 3. Distribution of fibrosis and myocardial disarray. ■: Fibrotic areas. ▨: Myocardium in disarray.

fibrosis were recognized together in the posterior part of the ventricular septum and the anterior wall of the left ventricle. These lesions extended from the epicardial aspect. Hypertrophic myocardial fibers were scattered in the fibrotic areas (Fig. 3). (2) Small patches of fibrosis were scattered in the wall of both ventricles and ventricular septum (Fig. 4). Papillary muscles of both ventricles also showed patchy fibrosis. (3) Fibrosis among the bundles of myocardial fibers was recognized more or less diffusely in the wall of both ventricles (Fig. 5). The ventricular septum also showed prominent fibrosis among myocardial bundles. (4) Fibrosis among the myocardial fibers was recognized near the scar-like fibrotic areas and small patchy fibrotic foci (Fig. 6). Slight fibrosis among myocardial fibers was observed in the areas with fibrosis among myocardial bundles.

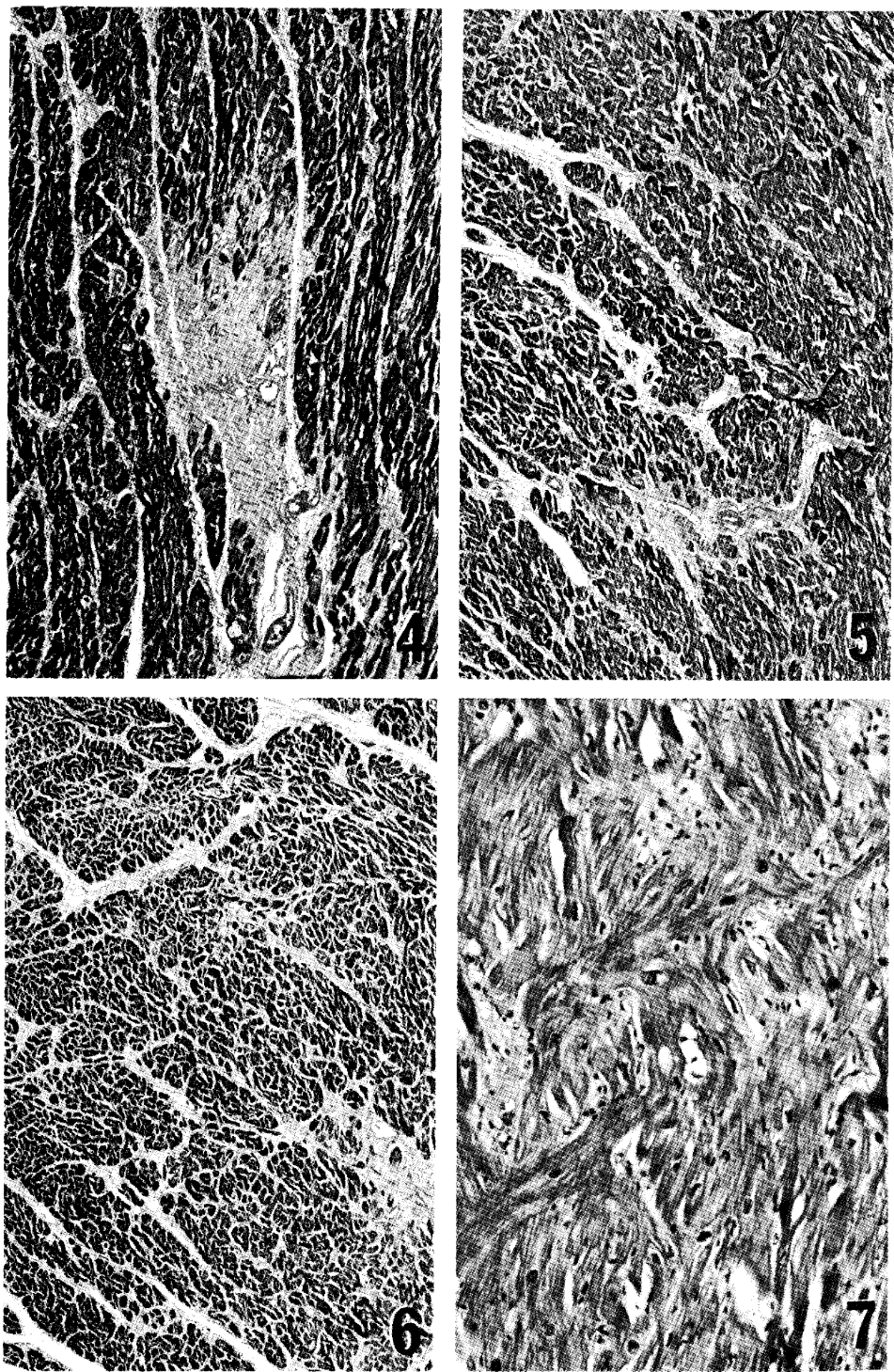
Hypertrophy of myocardial fibers was observed diffusely in the wall of both ventricles and the ventricular septum. The ventricular septum, posterior wall of both ventricles and antero-lateral wall of the left ventricle showed an interlaced arrangement of bizarre-shaped multipolar myocardial fibers, that is, so-called "disarray" (6) of hypertrophic myocardial fibers (Fig. 7). This disarray tended to appear in the areas adjoining scar-like fibrotic areas or patchy fibrotic foci (Fig. 3). The myocardial fibers beneath the endocardium and those in the papillary muscles of both ventricles generally showed pale swelling. Some areas of the ventricular septum also showed pale swelling of myocardial fibers. The endocardium showed no remarkable changes in general, although slight fibrotic thickening was observed

Fig. 4. A patchy fibrotic focus in the posterior wall of the left ventricle. (Masson's trichrome stain, $\times 40$)

Fig. 5. Fibrosis between myocardial bundles in the posterior wall of the left ventricle. (Masson's trichrome stain, $\times 30$)

Fig. 6. Fibrosis between myocardial fibers in the posterior wall of the left ventricle. (Masson's trichrome stain, $\times 30$)

Fig. 7. Disarrayed hypertrophic myocardial fibers in the ventricular septum. (HE, $\times 140$)



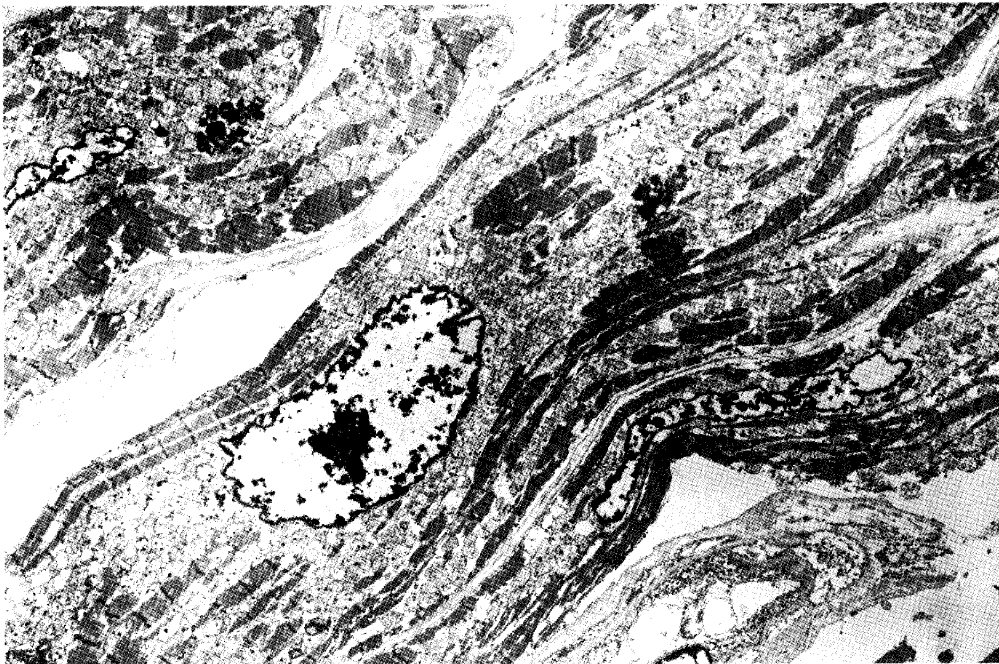


Fig. 8. Electron microgram of the myocardial fibers of the ventricular septum, showing mitochondrial hyperplasia and a decrease in myofilaments. ($\times 2,100$)

in some parts. The interstitium of the wall of both ventricles and ventricular septum showed minimal infiltration by lymphocytes.

Coronary arteries showed mild atherosclerosis and no stenotic changes. Intramyocardial arterioles showed no remarkable stenotic changes, although some arterioles in the scar-like fibrotic areas showed thickening of their intermediate layer. The base of a thrombus of the right atria was beginning to be organized. In the atrial wall, hypertrophy and pale swelling of myocardial fibers and fibrosis among myocardial fibers were observed. The sinoatrial node and atrioventricular node showed no prominent changes. Electron microscopic examination of the ventricular septum demonstrated a decrease in myofilaments and hyperplasia of mitochondria (Fig. 8). These changes were interpreted as corresponding to the microscopic pale swelling of myocardial fibers.

We diagnosed this case as HCM with ventricular dilatation from the above autopsy findings. Extensive myocardial hypertrophy with disarrayed arrangement, typical of usual HCM, was observed, and the family history of a daughter being clinically diagnosed as having HCM was also helpful in making the diagnosis. The direct cause of the ventricular dilatation was thought to be myocardial loss in the fibrotic areas.

DISCUSSION

Several possible causes of the usual HCM which progresses into the DCM-like state have been discussed in the literature (1, 2). In some cases (1, 2), loss of myocardial fibers and the resulting ventricular dilatation were attributed to stenosis of intramyocardial arterioles. However, in this case and also in some reported cases (4, 5), coronary arteries and intramyocardial arterioles did not show enough stenotic changes to explain the loss of myocardial fibers and fibrosis. Nimura *et al.* (4) stressed that the distribution of myocardial loss and fibrosis was closely related to that of disarrayed hypertrophic myocardial fibers. This tendency was clearly demonstrated in this case also. It might be that hypertrophic myocardial fibers in such HCM cases are more susceptible to relative oxygen deficiency or other unknown factors and tend to disappear, causing fibrosis. The distribution of myocardial loss and scar-like fibrosis in this case was somewhat different from that in the usual myocardial infarction, and the fibrosis of the scar-like areas was milder than in the usual myocardial infarction. These findings imply that the changes progressed slowly, and that the first change might be the gradual degeneration of myocardial fibers in relation to myocardial disarray. Considering these points, occurrence of familial DCM-like HCM (1, 5) might be explained from a genetic susceptibility of hypertrophic myocardial fibers. The possibility of myocarditis participating in such progression is sometimes mentioned (1, 4, 5). The heart of this patient at autopsy showed too mild an infiltration by inflammatory cells to consider the possibility of myocarditis. However, the absence of inflammatory cells in autopsy material cannot deny the post-myocarditic state. It is interesting that a case of post-myocarditic HCM was reported that showed dilatation of the ventricles and congestive heart failure later in the course (7). Myocarditis has to be left as one of the possible causes of DCM-like HCM, for patchy loss of myocardial fibers and fibrosis might suggest a post-myocarditic state.

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