A Case of Dilated Cardiomyopathy With Abnormal Atrioventricular Conduction 5 Years Before the Appearance of Left Ventricular Systolic Dysfunction

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The first manifestation of dilated cardiomyopathy is usually dyspnea as a result of left heart failure. In this report we describe an unusual patient with third-degree atrioventricular (AV) block unaccompanied by left ventricular dysfunction. Severe left ventricular dysfunction occurred 5 years after the implantation of a permanent pacemaker. We conclude that AV block should not be overlooked as an initial clinical manifestation of dilated cardiomyopathy.

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Key Words: Dilated cardiomyopathy; Atrioventricular block; Permanent pacemaker

Dilated cardiomyopathy is essentially a disease of the myocardium that is sometimes accompanied by conduction abnormalities. The incidence of atrioventricular (AV) block is approximately 20%, although the incidence of third-degree AV block is less than 10%.^{1,2} In general cases of dilated cardiomyopathy, left ventricular systolic function is already disturbed at the onset of AV block. We describe an interesting case of dilated cardiomyopathy that began with syncope and which was probably due to advanced AV block unaccompanied by left ventricular systolic dysfunction.

Case Report

A 39-year-old Japanese man was referred to our hospital for syncope. He had experienced episodes of syncope since the age of 21. At the age of 27, he had been referred to a private clinic for the evaluation of syncope and general malaise. At that time, a 12-lead electrocardiogram (ECG) showed complete left bundle branch block and first-degree AV block. At the age of

34, he was referred to our hospital because of a similar episode of syncope. At that time, the ECG showed third-degree AV block. A dual-chamber pacemaker was then implanted. The patient recovered and was discharged. He was observed thereafter at our hospital outpatient clinic.

Before his second admission, the patient complained of palpitations. An ECG showed paroxysmal atrial fibrillation, and the pacing mode was changed to VDD mode. He was readmitted to our hospital because of the sudden onset of left hemiplegia and speech disturbance. On admission (29 August 1995), his height was 160 cm, body weight 51 kg, blood pressure in the supine position 110/70 mmHg, and his heart rate was regular at 60 beats/min. Physical examination revealed a moderate holosystolic murmur (Levine 2/6) at the apex suggestive of mitral valve regurgitation. However, there was no sign of left heart failure. The liver was palpable 1 cm below the right costal margin in the mid-clavicular line. Tendon reflexes were present but diminished on the left side, and left-sided Babinski signs were noted.

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Fig 1. Electrocardiograms taken at a private clinic (top), and on first admission (middle) and second admission (bottom) to our hospital. Top: A complete left bundle branch block and first-degree AV block (PQ interval 0.22 sec). Middle: On the second admission the ECG on DDD mode pacing showed complete AV block. The QRS duration was slightly prolonged (0.20 sec) on the second admission (bottom), compared with the first admission (0.17 sec).

Laboratory tests indicated a slight elevation of serum creatinine kinase, which had normalized by the next day. Levels of antimyocardial antibody, and viral antibodies, such as anti-coxsackievirus and anti-echovirus antibodies, were not significantly elevated. Furthermore, ophthalmoscopic examination showed no abnormalities, and serum levels of angiotensin-converting enzyme and lysozyme were normal. Thallium-201 myocardial scintigraphy revealed small areas of hypoperfusion at the apex and on the

posterior wall. Chest radiographs showed a normal cardiothoracic ratio at the first admission (51%), and an increase at the second admission (63%). Serial 12-lead ECGs are shown in Fig 1.

When the patient was 27 years old, the ECG at the private clinic showed a complete left bundle branch block and first-degree AV block (PQ interval 0.22 sec). On the second admission, the QRS duration was slightly prolonged (0.20 sec) compared with the first (0.17 sec), although these QRS durations were on

Fig 2. Echocardiograms obtained at first admission (left) and second admission (right). The left ventricular end-diastolic dimension at the first admission was 4.5 cm. It had increased to 7.5 cm at the second admission. Left ventricular wall motion had deteriorated by the time of the second admission.

Fig 3. Light microscopic findings of right ventricular endomyocardial biopsy at first admission (left) and second admission (right) (H&E, 200 $\times$). On the first admission there is mild interstitial fibrosis and moderate disarrangement of the myocytes. On the second admission moderate interstitial fibrosis and cardiac myocyte degeneration is apparent.

pacing. On echocardiography, the left ventricular end-diastolic dimension at the first admission was 4.5 cm. It had increased to 7.5 cm at the second admission (Fig 2). The percentage fractional shortening gradually decreased from 34% at the first admission, to 26% after 2 years, 20% after 4 years, and finally to 14% at the second admission.

Histological examination of a biopsy specimen of the right ventricular endomyocardium obtained at the first admission showed very mild interstitial fibrosis and slight disarrangement of the myocytes. On the second admission, however, both the interstitial fibrosis and

degeneration of cardiac myocyte showed moderate increases (Fig 3). Based on the patient's clinical course and pathologic findings, we made a diagnosis of dilated cardiomyopathy.

After admission, computed tomography of the brain showed a low-density zone in the right cerebellum. It was treated as a cerebellar thromboembolism. The patient improved after 3 months' rehabilitation.

Discussion

In the present case, the onset of AV block occurred

while left ventricular systolic function was still completely preserved, and the patient has been observed for 5 years since the implantation of a DDD-mode permanent pacemaker, during which time systolic function has gradually deteriorated. Clinicians rarely have the opportunity to observe dilated cardiomyopathy in its latent stage. We therefore consider that this case is important in evaluating the clinical history of dilated cardiomyopathy.

Our patient had experienced several episodes of syncope since the age of 21. The first symptoms of dilated cardiomyopathy are usually caused by heart failure, eg, dyspnea.³ The incidence of AV block in these patients is approximately 20%^{1,2} and heart failure is usually also present. This patient showed AV block as an initial symptom without heart failure. The cause of syncope in this case could be arrhythmia, such as the arrhythmia of sick sinus syndrome, ventricular tachycardia. However, we would like to believe that in this case syncope was caused by advanced AV block.

Sekiguchi et al⁴ proposed the existence of a group of diseases, which they called arrhythmia-conduction (electric) disturbance-type cardiomyopathy (ECM), and described 4 features of the disease: (1) the presence of AV block, complete right bundle branch block, left bundle branch block, intraventricular conduction delay, ventricular arrhythmia or atrial arrhythmia; (2) the presence of significant pathological changes in the right ventricle, left ventricle, and/or right endomyocardial biopsy specimen; (3) a non-hypertrophic, non-dilated form; (4) the absence of significant changes on the coronary arteriogram. Obviously, the findings in our case differ from those of ECM. As we have no data regarding the pathology of the left myocardium, the findings of the right-sided endomyocardial biopsy can be taken to represent those occurring on the left side.⁵ There were no significant pathological findings on the first endomyocardial biopsy and normal left ventricular wall motion on echocardiography, even although third-degree AV block was apparently present. We therefore consider that this patient may belong to a slightly different category.

Regarding the pathological findings of the right ventricular endomyocardial biopsy specimen, minor interstitial fibrosis and disarrangement of myocytes were observed in the first biopsy. It would be difficult to diagnose dilated cardiomyopathy on the basis of these histological changes alone. However, the changes found on the second biopsy and the patient's clinical course make it possible to diagnose dilated cardiomyopathy in this case. If we could observe the pathological findings in the latent stage of dilated

cardiomyopathy, then this case could be important. Compared with patients with postmyocarditis, our patient did not exhibit significant inflammatory changes on first admission. Considerable interstitial fibrosis and cardiac myocyte degeneration were noted in the second biopsy, therefore myocarditis was unlikely.

The insertion of a pacemaker lead into the right ventricle may cause an adverse reaction in the right ventricular myocardium. So-called metal allergy has been reported, and pacemakers sometimes cause contact dermatitis or an allergic reaction to metal in a pacemaker pocket.⁶ Pacemaker electrical stimulation may cause uncoordinated myocardial activation, resulting in the disarray of myocardial fibrils.⁷⁻¹⁰ Although we did not perform a left ventricular endomyocardial biopsy in this patient, left ventricular disturbance was obvious on echocardiography. We cannot completely exclude permanent pacing as a cause of left ventricular dysfunction in this case.

In conclusion, we emphasize the existence of a group of dilated cardiomyopathies in which the initial symptom is a conduction abnormality followed by a gradual deterioration of cardiac function.

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