CASE REPORTS

An Isolated Left Ventricular Lesion Associated with Left Ventricular Tachycardia

Arrhythmogenic “Left” Ventricular Dysplasia?

Masanori Okabe, M.D., Keisuke Fukuda, M.D.
Yoshiyuki Nakashima, M.D., Kuikuo Arakawa, M.D.
and Masahiro Kikuchi, M.D.*

We present a patient who had a localized myocardial lesion of the left ventricle. Major clinical sequelae were left ventricular tachycardia and heart failure. This case appears to represent a left-sided counterpart of arrhythmogenic right ventricular dysplasia.

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RIGHT ventricular (RV) myopathy associated with RV tachycardia is referred to as arrhythmogenic RV dysplasia.1,2 We report a patient who had a localized non-ischemic lesion of the left ventricle (LV) associated with LV tachycardia and diffuse LV dysfunction. We speculate that the RV myopathy and arrhythmogenic potential seen in RV dysplasia1,2 might also apply to the LV disease in the present case.

CASE REPORT

A Japanese man who had been asymptomatic until age 55, developed palpitation episodes of which lasted several minutes each and occurred up to once a week. He experienced two episodes of syncope when aged 55 and 57 years, both of which required electrical cardioversion for termination of sustained ventricular tachycardia (VT). In the first episode, VT was transformed into ventricular fibrillation after the intravenous infusion of lidocaine. Thereafter, ventricular arrhythmia was controlled by disopyramide, but administration of this drug was discontinued prior to the second episode because the patient relocated. At age 58, the frequency and duration of palpitation episodes increased and symptoms, including weakness and lightheadedness during the episodes, worsened. The patient was then referred to our hospital for evaluation of cardiac disease.

There was no family history of neuromuscular disease. The patient denied symptoms suggestive of congestive heart failure. He was 162 cm tall and weighed 63 kg. The pulse was 67 per min and regularly irregular. The blood pressure was 110/78 mmHg. A fourth heart sound and a grade-three holosystolic murmur were audible. Neither organomegaly nor peripheral edema was observed. Neurological examination was normal. Laboratory findings showed a mildly positive CRP (± to 2+) and impaired glucose tolerance, which established the a diagnosis of diabetes mellitus. The cardiothoracic ratio (CTR) was 61% on a chest radiograph (Fig. 1). The resting electrocardiogram (ECG) showed sinus rhythm with a slightly prolonged QRS duration. Small q waves were noted in the inferior leads. Ambulatory ECG monitoring documented

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Departments of Internal Medicine and *Pathology, Fukuoka University School of Medicine, Fukuoka, Japan
Mailing Address: Masanori Okabe, M.D., Department of Internal Medicine, Fukuoka University School of Medicine, 7-45-1 Nanakuma, Jonan-ku, Fukuoka 814-01, Japan

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Fig. 1. Chest radiograph and electrocardiograms.
The chest radiograph demonstrates a cardiothoracic ratio of 61% (left panel). The upper right shows a twelve-lead recording obtained during sinus rhythm. The lower right was recorded during the an episode of ventricular tachycardia at age 55.

Fig. 2. Ambulatory electrocardiographic monitoring. Incessant ventricular tachycardia is shown by ambulatory ECG monitoring.

multiform premature ventricular contractions and incessant VT (Fig. 2). VT was monomorphic in a right bundle branch block pattern with left axis deviation (Fig. 1). Cardiac catheterization revealed that the pulmonary arterial pressure, mean pulmonary wedge pressure, and LV pressure were 25/12, 11, and 130/10 mmHg, respectively. Left ventriculography demonstrated diffusely hypokinetic wall motion and a nearly akinetic posterior segment with grade-1 mitral regurgitation (Fig. 3). Although the LV ejection fraction (23%) was markedly reduced, the cardiac index (thermodilution method: 3.20 L/min/m²) was maintained by an increased LV volume (end-diastolic volume index; 152 mL/m²). Coronary arteriography was normal (Fig. 4). Myocardial dysfunction could not be attributed to diabetes mellitus, since the serum glucose concentration was well-controlled by diet therapy, and there was no evidence of dia-
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Fig. 3. Left ventriculography.
A, B and C, D are diastolic and systolic images of the left ventriculography in the right anterior oblique view and the left anterior oblique view, respectively. These images were obtained during sinus rhythm without administration of cardiovascular therapeutics.

Fig. 4. Coronary angiography.
A and B: Angiography of the right and left coronary arteries, respectively, in the left anterior oblique view.

abetic microangiopathy. Therefore, we concluded that the diagnosis was idiopathic dilated cardiomyopathy. Holter ECG assessment suggested the efficacy of atenolol and mexiletine against ventricular arrhythmias.

For the following two years, the patient was free of cardiac events on oral atenolol (100 mg/day), except for infrequent palpita-
tion episodes, with a stable CTR (from 60 to 62%) and echocardiographic parameters (LV diastolic dimension; from 59 to 62 mm, fractional shortening; from 15 to 20%). However, the patient developed intolerance to atenolol because of malaise and chest discomfort. The daily dose of atenolol was decreased over one and a half years. mexiletine (300 mg/day) was then prescribed, since palpitation episodes became frequent. Three months after initiating mexiletine, heart failure became overt. The patient was hospitalized and given oral diuretics. The CTR improved with therapy (from 69 to 60%), but the echocardiographic LV diastolic dimension did not change significantly (70 mm). Six months later, the patient was readmitted to our hospital because of intractable heart failure which was eventually complicated by fatal cerebral infarction at age 63. Thus, dysfunction of the LV and recurrent VT had been significant problems through- out the clinical course.

The heart at autopsy weighed 470 g and showed a dilated LV. A translucent area of marked wall thinning was observed in the posterior wall of the LV (Fig. 5). Histopathologic examination of the lesion showed transmural fibro-fatty replacement of myocardium (composed mainly of fatty tissue) with some infiltrating lymphocytes (Fig. 6). This myocardial lesion was likely the origin of VT, as determined by the QRS morphology. Other portions of the LV were nearly normal in wall thickness and showed only mild interstitial fibrosis. The RV was morphologically normal. No mural thrombi were found in any cardiac chamber. No significant luminal narrowing of the epicardial coronary arteries was observed.

DISCUSSION

Focal myocardial lesions can usually be attributed to identifiable diseases, such as ischemic heart disease or myocarditis. Although we cannot eliminate the possibility of coronary embolism in the present case, the lesion was not consistent with a healed myocardial infarct. The extent of the lesion did not correspond to the distribution of the major epicardial coronary arteries. The posterior descending and posterolateral branches of the right coronary artery diverged along the edge of the lesion (Fig. 5). There was no pathologic evidence of myocarditis. Some patients with heredofamilial neuromuscular disorders also exhibit cardiomyopathy with focal myocardial myocardial lesions. However, the present case had no abnormalities in the skeletal muscle, and did not show tall right precordial R waves with deep limb lead, which are ECG characteristics of progressive muscular dystrophy. Although heterogeneity of the extent of the myocardial lesions is not uncommon in patients with dilated cardiomyopathy, transmural ventricular lesions are rare. In a review of 152 necropsy cases of dilated cardiomyopathy, 22 had a focal “ventricular scar”, but none showed a “transmural scar” of the ventricular wall. In addition, these grossly visible scars appeared fibrous rather than as fatty replacement on histologic examination.

Focal LV lesions similar to that of our
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Fig. 6. Pathologic findings in the thinned portion of the left ventricle.
A: A horizontal section of the ventricles. The left ventricle shows severe dilatation and extraordinary thinning of the posterior wall (arrows), which is only 1.5-mm thick. B: Opened left ventricle, exposing the aortic valve and aorta. The posterior papillary muscle (arrows) is extensively involved. C: Transmural fibro-fatty replacement observed in the posterior wall of the left ventricle. Masson's trichrome stain; original magnification ×5. The left ventricular cavity is oriented to toward the bottom. D: Residual myocardial cells, darkly stained by an immuno-histochemical stain for myoglobin, are seen only in the trabecular region. Original magnification ×5. E: A higher magnification shows scattered lympho-cytic infiltration. Hematoxylin-eosin stain; original magnification ×132.
lt: left, ant: anterior.

Patient have been described in two specific conditions: in Chagas’ disease \(^7,8\) and in RV dysplasia and allied diseases? Patients with Chagas’ disease present with ventricular arrhythmias and a dilated failing heart with focal myocardial lesions. Morphologically, marked thinning of the ventricular wall is commonly observed at the ventricular apex \(^7,8\). The disease is prevalent in Central and South America, although it may rarely be found in nonendemic areas as a consequence of transfusion with contaminated blood products. Our patient had many clinicopathologic features similar to cases of those in Chagas’ disease, but the patient had never been abroad or received transfusions.

The clinical features of RV dysplasia are characterized by the presence of ventricular postexcitation waves and recurrent VT with a QRS configuration of left bundle branch block (arrhythmogenic RV dysplasia). Angiocardiography typically demonstrates a dilated and diffusely hypokinetic RV with localized areas of dyskinesis. The pathologic hallmark of the disease is lipomatosis of the RV free wall, usually seen as localized wall thinning \(^1,2\). Furthermore, RV dysplasia is believed to exclusively involve the RV. However, some reports have demonstrated functional \(^10,11\) and pathologic \(^9,12\) abnormalities of the LV in patients with RV dysplasia. Prospective radionuclide angiocardiographic studies \(^10\) revealed global and segmental LV dysfunction during exercise in all 6 of the patients with arrhythmogenic RV dysplasia. These findings suggest that RV dysplasia might actually be a generalized myocardial disease with predominant RV involvement.
In the present case, recurrent LV tachycardia and heart failure were major clinical problems. Left ventriculography showed generalized hypokinesis with focal akinesis. Furthermore, the localized thinned portion of the LV indeed contained fatty tissue. We speculate that the present case might represent a left-sided counterpart of RV dysplasia.

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REFERENCES