Case Reports

Familial Dilated Cardiomyopathy Complicated by Left Ventricular Aneurysm

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SUMMARY

Two siblings presented with symptoms of left ventricular dysfunction and ventricular arrhythmias. Echocardiography and left ventriculography revealed dilatation, diffuse hypokinesis and apical aneurysm of the left ventricle in both cases. Myocardial infarction was unlikely by history and examinations. We diagnosed them as cases of familial dilated cardiomyopathy complicated by left ventricular aneurysm.

Additional Indexing Words:

Dilated cardiomyopathy Left ventricular aneurysm

DILATED cardiomyopathy with left ventricular aneurysm has been rarely reported,^{1,2)} and there are no reports of familial occurrence of this association. We report herein two siblings with dilated cardiomyopathy complicated by left ventricular aneurysm.

CASE REPORT

Case 1:

A 56-year-old man was admitted to our clinic because of the gradual onset of chest discomfort and dyspnea lasting for a few hours. He had been suffering from the symptoms for 14 years, but had no history of chest pain suggestive of ischemic heart disease or myocarditis.

On admission, an electrocardiogram (ECG) showed sustained ventricular tachycardia. After electrical cardioversion, ECG showed Q waves in leads III, aV_F, and V₁-V₃, diffuse ST-T abnormalities, complete left bundle branch block, and first-degree atrioventricular block (Fig. 1). Frequent,

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Received for publication August 25, 1989.

Accepted October 4, 1989.

Fig. 1. A: Electrocardiogram of case 1 showing complete left bundle branch block. B: Holter ECG of case 1 showing frequent, multifocal ventricular premature beats.

multifocal ventricular premature beats and short runs of ventricular tachycardia were found on a Holter ECG. Physical examination revealed third and fourth heart sounds, a mild regurgitant systolic murmur at the apex, and bubbling rales in the bilateral lower lung fields. Laboratory data including cardiac enzymes gave normal results. A chest radiogram showed cardiomegaly with a cardiothoracic ratio of 67 percent, and signs of pulmonary congestion. First-pass radionuclide angiography by ^{99m}Tc -pertechnetate revealed a decreased ejection fraction of both ventricles (left, 22%; right, 23%). ^{201}Tl scintigraphy at rest showed a persistent perfusion defect at the anteroapical wall of the left ventricle. Echocardiography and left ventriculography disclosed marked dilatation, diffuse hypokinesis, and apical aneurysm of the left ventricle, and mild mitral regurgitation. Coronary arteriography showed normal coronary arteries (Fig. 2). He was treated with mexiletine and metoprolol for ventricular tachycardia, and digoxin and diuretics for congestive heart failure. However, attacks of ventricular tachycardia and ventricular fibrillation with congestive heart failure have repeatedly occurred.

Case 2:

A 59-year-old man, an elder brother of case 1, was hospitalized because of exertional dyspnea and attacks of chest discomfort. He had been suffering from the symptoms for 15 years. He had no history of chest pain suggestive of angina pectoris or myocardial infarction. On admission, physical exami-

Fig. 2. A: Left ventriculograms in the right anterior oblique projection in diastole (a) and systole (b) of case 1. Arrows indicate left ventricular aneurysm in the apical wall. B: Right (a) and left (b) coronary arteriograms of case 1 show normal coronary arteries.

Fig. 3. A: Electrocardiogram of case 2 showing complete left bundle branch block. B: Holter ECG of case 2 showing short runs of ventricular tachycardia.

nation revealed a fourth heart sound and a mild regurgitant systolic murmur at the apex. A chest radiogram showed a cardiothoracic ratio of 57 percent. ECG showed Q waves in leads V₁–V₃, diffuse ST-T abnormalities, complete left bundle branch block, and first-degree atrioventricular block (Fig. 3). Frequent, multifocal ventricular premature beats, and short runs of ven-

tricular tachycardia were found on a Holter ECG. First-pass radionuclide angiography by ^{99m}Tc -pertechnetate revealed impaired left ventricular function and preserved right ventricular function with ejection fractions of 25 and 57 percent, respectively. ^{201}Tl scintigraphy at rest showed a persistent perfusion defect at the anteroapical wall of the left ventricle. Echocardiography and left ventriculography disclosed dilatation and diffuse hypokinesis of the left ventricle with apical aneurysm, and mild mitral regurgitation. Coronary arteriography showed normal coronary arteries (Fig. 4). He was treated with mexiletine and enalapril, but attacks of ventricular tachycardia and congestive heart failure have recurred.

Examination of the ECGs and echocardiograms of 11 of their relatives (their mother, 1 sister and 2 brothers, 3 children, and 4 nephews) revealed no abnormality suggestive of cardiomyopathy.

DISCUSSION

Our patients were two siblings presenting with almost the same signs and symptoms of left ventricular dysfunction and arrhythmias. Left ventriculography revealed diffuse hypokinesis and apical aneurysm of the left ventricle in both cases (Figs. 2 and 4). Although the most common cause of left ventricular aneurysm in adults is myocardial infarction,³⁾ it was unlikely

Fig. 4. A: Left ventriculogram in the right anterior oblique projection in diastole (a) and systole (b) of case 2. Arrows indicate left ventricular aneurysm in the apical wall. B: Right (a) and left (b) coronary arteriograms of case 2 show normal coronary arteries.

in our cases because they had no history of chest pain suggestive of angina pectoris or myocardial infarction and coronary arteriography showed normal coronary arteries (Figs. 2 and 4). Coronary embolism might account for ventricular aneurysm but does not explain diffuse hypokinesis of the left ventricle. They had never experienced chest trauma which might cause aneurysm and diffuse hypokinesis. Cases of congenital left ventricular aneurysm have been reported,⁴⁾ and we could not exclude this possibility completely. Because we could not find any clear etiology of dilatation and diffuse hypokinesis of the left ventricle in our cases, we diagnosed them as idiopathic dilated cardiomyopathy.

Dilated cardiomyopathy is rarely complicated by left ventricular aneurysm.^{1),2)} No report has described familial cases of the complication, although there are some reports of familial dilated cardiomyopathy.^{5),6)} A previous report suggested that left ventricular aneurysm was associated with transmural myocardial fibrosis which might be related to a basic pathologic abnormality responsible for the development of dilated cardiomyopathy.²⁾ In our cases, transmural fibrosis might exist where left ventricular aneurysm was present because persistent perfusion defects in that area were found by myocardial perfusion scintigraphy. It is conceivable that the left ventricular aneurysms in our familial cases may be associated with a basic pathologic abnormality of dilated cardiomyopathy.

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