Histological Abnormalities in a Biopsy Specimen Obtained from the Left Ventricle without Reduced Function in a Patient with Right Ventricular Dysplasia

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Left ventricular histological examination was performed in a patient with right ventricular (RV) dysplasia. Although the left ventricle (LV) appeared to have a normal shape and function, the LV biopsy specimen revealed apparent myocyte hypertrophy and substantial fibrotic changes without fatty infiltration. It was not clear whether these considerable histological changes were part of right ventricular dysplasia or not. The present case indicates that even with normal LV shape and function in patients with right ventricular dysplasia, histological examination of LV may provide additional information on its pathogenesis and prognosis.

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Introduction

Right ventricular dysplasia (RVD) is characterized by an abnormality in the development of part of the right ventricular musculature. The right ventricular musculature is replaced by adipose or fibroadipose tissue (1, 2). RVD is frequently associated with ventricular tachycardia of left bundle branch block configuration. Therefore, there are many clinical reports (3, 4) with respect to the mechanism and treatment of this life-threatening tachycardia.

Interestingly, recently much attention has been focused on left ventricular involvement in RVD, with regard to the etiology and prognosis of this disease. Initially, it was thought that the pathological changes were limited to the right ventricle (1, 2, 5). Recently, a few studies (2, 5-7) demonstrated left ventricular abnormalities on echocardiography or ventriculography. However, there have been only 3 cases in which histological examination was performed on the left ventricle with normal shape and function (5).

Here, we report the histological findings of a left ventricle and normal shape and function.

Case Report

Case

A 66-year-old female was admitted with a 2-year history of mild chest pain and discomfort. She had never had symptoms of palpitations or faintness. Her past and family histories revealed no particular finding. On admission, her blood pressure was 110/60 mmHg with heart rate of 60 bpm. There was no audible heart murmur. She had had no episode of right-sided heart failure. Chest roentgenogram showed an enlarged cardiac shadow with a cardiothoracic ratio of 63%. Conventional 12-lead electrocardiogram revealed a characteristic post-excitation wave (i.e., epsilon wave) in leads V1 to V3 (Fig. 1).

Echocardiogram

Two-dimensional echocardiogram revealed marked right ventricular (RV) dilatation with diffuse reduced wall motion, and in particular, a dyskinetic bulge of the RV inflow tract. In contrast, the left ventricle (LV) appeared to be of normal size and function, with an LV diastolic dimension of 42 mm and ejection fraction of 63% with no localized hypokinetic lesion (Fig. 2).

Catheterization

Neither abnormal pressure nor oxygen saturation was found on right heart catheterization. Coronary angiogram showed normal coronary arteries. Right ventriculogram demonstrated diffuse wall motion reduction, bulging formation at the inflow tract and marked dilation of the outflow tract (Fig. 3). In contrast, the left ventriculogram showed normal wall motion.

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Fig. 1. A 12-lead electrocardiogram on admission. Post-excitation wave (i.e., epsilon wave) in leads V1 to V3 is noted (arrow in the right panel). Incomplete right bundle branch block with T-wave inversion in leads V1 to V4 are also noted.

with normal volume.

Electrophysiological study

Endocardial ventricular post-excitation waves were recorded at the inflow and outflow of the right ventricle, but ventricular tachycardia could not be induced by stimulating the right ventricle using the extrastimulus or burst technique. Post-excitation waves were also recorded from the body surface signal averaging electrocardiogram.

Histological findings

To confirm the diagnosis, biopsy was performed with informed consent. Both of the specimens obtained from the RV showed interstitial fat infiltration, hypertrophy and mild degeneration of myocytes (Fig. 4), which are consistent with RVD.

Four biopsy specimens were obtained from the lateral wall of the left ventricle. These specimens showed hypertrophy of myocytes with mild disarray and considerable fibrotic changes with myocyte degeneration (Fig. 5), which are not inconsistent with dilated cardiomyopathy. Although these changes were mild in one sample, the findings were essentially the same in the other 3 specimens. Lymphocytic infiltration was not observed in the right or the left ventricular biopsy specimens.

Discussion

In the present RVD case, substantial abnormal histological findings in the LV biopsy specimen were observed without reduced function and enlargement of the LV.

The diagnosis of RVD was made in the present patient. Marked dilation of right ventricle and bulging formation of the right ventricular inflow tract were consistent with the characteristic findings of RVD. Although the degree of fatty infiltration seemed to be moderate compared to the typical RVD right ventricle, the histological findings were consistent with those of RVD. Therefore, the diagnosis of RVD in the present case seemed reasonable.

Although the echocardiogram and ventriculogram revealed a normal LV shape and function in this patient, the left ventricular biopsy specimen showed obvious histological changes without fatty infiltration, which is not inconsistent with the findings of idiopathic dilated cardiomyopathy. Blomstrom-Lundqvist et al (8) examined 13 RVD patients and detected left ventricular hypofunction and/or enlargement by cineangiography in 5 patients. Similarly, Pinamonti et al (5) reported that 14 of 39 RVD patients already had LV abnormalities on their first examination, and dilation and/or hypofunction of LV was observed during the 7-year follow-up period in 6 of the remain-
Fig. 2. Echocardiographic findings. Long-axis (A) and short-axis (C) two-dimensional echocardiogram shows marked enlargement of the right ventricle (RV). M-mode echocardiogram (B) shows non-dilated and normal function of left ventricle (LV). Inter-ventricular septal motion was not paradoxical. Four-chamber view (D) revealed bulge formation of right ventricular inflow wall (arrows). RA: right atrium, LA: left atrium.

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ing 22 RVD patients without LV abnormalities at their first examination. Although it was unclear in the present case whether these histological changes were part of right ventricular dysplasia or not, these reports and the present case suggest that RVD may involve the left ventricle in some instances.

In the present case, the left ventricular biopsy specimen showed apparent myocyte hypertrophy and considerable fibrotic changes without fatty infiltration. In most studies, left ventricular abnormalities have been evaluated using echocardiography, radionuclide angiography and/or left ventricular angiography; histological examination of the left ventricle was performed in only a few cases (5). Furthermore, most histological studies examined cases with reduced LV function and found variable histological findings, ranging from mild nonspecific findings to typical findings of RVD, including fatty infiltration. There have been only 3 cases reported in which LV histological examination was performed in RVD patients without reduced LV function (5). In these 3 cases, interstitial fibrosis, and hypertrophy and attenuation of myocytes were observed. These 3 cases, in addition to the present case, indicate that LV histological abnormalities may be present even in patients without reduced function and this fact may be important with respect to the prognosis.

The reason for the discrepancy between the normal LV function and histological abnormal findings in this case was obscure. Similarly, no explanation for this discrepancy was found in the 3 cases discussed above. Because these findings were obtained in only 4 biopsy specimens and all of the 4 specimens were obtained from the lateral wall of the left ventricle, the extent of the histological abnormalities could not be determined. The distribution of these abnormalities may be limited. Although notable fibrotic changes were observed, the degeneration of myocytes appeared to be mild and associated without reduced ventricular function.

In summary, left ventricular histological examination may provide additional information on the pathogenesis and prognosis of patients with RVD.
Fig. 3. Right ventricular angiogram of diastolic phase. Localized akinetic large bulge at inflow wall is observed on anterior view (A). Remarkable dilatation of outflow tract is also observed on lateral view (B).

Fig. 4. Microscopic findings of biopsy specimen obtained from the right ventricle. Fatty infiltrations are noted (HE stain, A: x200, B: x400).

References

Fig. 5. Microscopic findings of biopsy specimen obtained from the left ventricle. Hypertrophied myocyte with mild disarray and considerable fibrotic changes are noted. A) HE stain (x100), B) Azan-Mallory stain (x100), C) HE stain (x400), D) Azan-Mallory stain (x400).