A CASE OF CONGENITAL LEFT VENTRICULAR DIVERTICULUM WITH PULMONARY STENOSIS AND ITS SCINTIGRAPHIC CHARACTERISTICS

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We encountered a 31-year-old female patient with mild valvular pulmonary stenosis who had no abnormality in the electrocardiogram but pulmonary dilatation in the chest radiograph. Two-dimensional echocardiography and magnetic resonance imaging demonstrated two small protrusions at the interventricular septum indicating diverticula. Large perfusion defects were observed at the anterior wall in the thallium-201 myocardial tomograms. Short axial and vertical long axial images by ECG-gated blood pool tomography revealed an outpouching best seen during diastole and a good contraction during systole in the corresponding areas. These findings suggested the presence of thin but normal myocardium in the anterior wall, i.e., a muscular type of left ventricular diverticulum. The presence of the muscular type of left ventricular diverticulum at the anterior and septal walls was confirmed by contrast left ventriculography. A congenital diverticulum at the anterior and septal walls with pulmonary stenosis is very rare. Furthermore, its scintigraphic images were quite characteristic and useful for its diagnosis.

A congenital left ventricular diverticulum is a relatively rare cardiac anomaly and has to be differentiated from other disorders showing saccular protrusion from the left ventricular contour such as an aneurysm or a localized herniation through a partial pericardial defect. Histologically, congenital diverticula are divided into two types, muscular and fibrous. Clinically, they can be differentiated by their contractile properties, i.e., the muscular type has normal contraction and the fibrous type has paradoxical motion (dyskinesis) during systole. Therefore, it is essential to evaluate the morphology and contraction pattern of the lesion in order to differentiate it from other diseases and diagnose the type of diverticulum. The diagnosis of left ventricular diverticulum has been made by two-dimensional echocardiography and contrast angiography. Recent advances in non-invasive methods such as radionuclide and magnetic resonance imaging are expected to contribute to the diagnosis of various cardiac anomalies. Although radionuclide techniques, especially three-dimensional imaging by single photon emission computed tomography (SPECT), have been established for assessing cardiac structure, function and coronary perfusion, there are very few scintigraphic observations of this disease.

Key words: Congenital left ventricular diverticulum, Pulmonary stenosis, Gated blood pool tomography, Thallium-201 myocardial tomography, Two-dimensional echocardiography

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In this report, we present a very rare case of a diverticulum at the interventricular septum and anterior wall with pulmonary stenosis and describe its scintigraphic characteristics revealed by the combined use of ECG-gated blood pool and thallium-201 myocardial tomographies.

CASE REPORT

The patient, a 31-year-old woman, was admitted to our department on February 9, 1988 for evaluation of her abnormal heart murmur. At the age of 6, a systolic heart murmur had been noted but there had been no further investigations. When she was admitted to the Department of Gynecology in our hospital for operation on a myoma uteri 25 years later, the heart murmur and an abnormality on chest roentgenogram were pointed out. Her family history revealed a consanguineous marriage and a cousin with ventricular septal defect. No anomalies in the thoracic or abdominal wall, nor jugular venous distention, were detected. Carotid pulsation was normal and physical examination showed no abnormality in heart rate nor blood pressure whereas a grade 3/6 of systolic murmur at the second left intercostal space near the sternum was auscultated. No hepatomegaly, peripheral edema nor neurological abnormality were detected. The electrocardiogram (ECG) showed biphasic T wave in the leads of V₂ and V₃ (Fig. 1). Her chest radiograph revealed 46% cardiothoracic ratio and abnormal prominence of the second left cardiac silhouette but no pulmonary venous congestion, suggesting dilatation of the pulmonary artery (Fig. 2).

Two-dimensional echocardiography revealed an outpouching at the interventricular septum which contracted during systole (Fig. 3). ECG-gated magnetic resonance imaging demonstrated 2 small protrusions at the interventricular septum (Fig. 4). Large perfusion defects were observed at the anterior wall in the thallium-201 myocardial tomograms (Fig. 5). Short axial and vertical long axial images by ECG-gated blood pool tomography revealed an outpouching best seen during diastole and a good contraction during systole in the corresponding areas (Fig. 6). These findings suggested a muscular type of left ventricular diverticulum.
Fig. 3. Two-dimensional echocardiogram from the long-axis view showing an outpouching at the interventricular septum which contracts during systole (arrows). RV = right ventricle, LV = left ventricle, RA = right atrium, LA = left atrium, Ao = aorta.

Fig. 4. ECG-gated magnetic resonance imaging; coronal (left), four chamber (middle) and sagittal views (right) at the end-systolic phase, showing outpouchings (arrows) at the septal and anterior walls. RV = right ventricle, LV = left ventricle, RA = right atrium, LA = left atrium, Ao = aorta, CS = coronary sinus.
A contrast left ventriculogram confirmed the presence of left ventricular diverticulum in the anterior and septal walls (Fig. 7). In addition, two-dimensional and Doppler echocardiography showed mild right ventricular hypertrophy, pulmonary dilatation and a mild pressure gradient (13 to 18 mmHg) between the main pulmonary artery and right ventricle, indicating the valvular type of pulmonary stenosis. However, left-sided hemodynamics were normal and no abnormal shunt flow was detected. Coronary angiography and endomyocardial biopsy from the right ventricular septum did not show any abnormalities.

**DISCUSSION**

Clinically, congenital left ventricular diverticulum has been classified as (1) a diverticulum associated with midline thoracoabdominal defects, (2) a fibrous diverticulum at an apical or subvalvular portion which can be seen in native Africans, (3) an isolated apical diverticulum without any thoracic wall anomalies and (4) single or multiple left ventricular diverticula along either the diaphragmatic or anterior ventricular wall. The diverticulum in our case is not one of the first three types above but may be one of the left ventricular diverticulosis reported by
Balatax et al. In our case, histological evidence of congenital diverticulum was not obtained and it was not determined whether or not the diverticulum was multiple. The diagnosis of a muscular type of congenital diverticulum was made based on characteristic morphology and contracting pattern during systole. Diverticulum at the septal wall is very rare and no cases with pulmonary stenosis have been reported in the previous literature.1–9 Rajpal et al.3 and Kato et al.5 reported a left ventricular diverticulum having normal uptake of thallium-201 at the lesion. However, there been no cases with left ventricular diverticulum showing perfusion defects on thallium-201 images in which normal contraction was confirmed. Perfusion defects with diastolic bulge can also be seen in a postinfarction ventricular aneurysm. In our case, however; the possibility was excluded because there was no abnormality in the electrocardiogram, normal contraction during systole and intact coronary angiograms. The mechanism of perfusion defects is not clear. We believe that these findings implicate the presence of 'thin but normal' myocardial layers at the lesion, i.e. a muscu-

Fig.6. ECG-gated blood pool images; planar image from modified left anterior oblique view (A), serial coronal tomograms from apex (bottom) to cardiac base (top)(B), and vertical long axial tomogram (C), demonstrating the outpouching at the anterior portion during diastole (arrows) and a good contraction during systole. RV = right ventricle, LV = left ventricle, RVOT = right ventricular outflow tract.

Fig.7. Right anterior oblique (left) and left anterior oblique (right) views of contrast left ventriculogram during end-diastole (top) and end-systole (bottom). Arrowheads indicate a left ventricular diverticulum which contracts during systole. ED = end-diastole, ES = end-systole, RAO = right oblique view, LAO = left oblique view.

lar type of ventricular diverticulum.

In diagnosing left ventricular diverticulum, it is essential to assess the regional wall motion because of the need for differentiation from aneurysm. In addition to conventional contrast left ventriculography, non-invasive cardiac imaging has contributed to detecting cardiac abnormalities. In particular, single-photon emission computed tomography (SPECT) has improved the diagnostic efficacy of scintigraphic imaging with its three-dimensional imaging ability. We have also reported on the morphological and functional assessment of regional cardiac abnormalities by SPECT in ischemic and congenital heart diseases. In our case, a systolic protrusion and a normal contraction were observed at the corresponding region to the perfusion defects by the gated blood pool SPECT. Thus, SPECT imaging was very useful for detecting left ventricular diverticulum by assessing the cardiac morphology and regional wall motion. However, this scintigraphic technique failed to delineate the form of the diverticulum in detail compared to contrast left ventricular angiography. This is probably because of its poor spatial resolution.

The clinical significance of congenital diverticulum reported here is not necessarily clear. Our patient has been asymptomatic and it is said that the muscular type of diverticulum produces no local nor systemic complications such as heart failure, angina pectoris, ventricular tachycardia, cardiac tamponade, infectious endocarditis nor systemic emboli as seen in the fibrous type. Thus, a congenital diverticulum at the anterior and septal walls with pulmonary stenosis not associated with any other anomalies appears very rarely and its scintigraphic image by SPECT was very characteristic and useful for diagnosis.

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