Congenitally Corrected Transposition of the Great Arteries in a 65-Year-Old Woman

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SUMMARY

A 65-year-old Japanese woman was admitted to hospital because of exertional dyspnea. Transthoracic echocardiography showed diffuse hypokinesis of the left-sided ventricular wall, but was not clear enough to provide useful information because of the rotation of the cardiac apex and the presence of lung tissue. Systemic ventriculography showed that the left-sided ventricle with heavy trabeculations was morphologically similar to a normal right ventricle. Magnetic resonance imaging (MRI) clearly revealed corrected transposition of the great arteries. Because this patient had no severe associated cardiac anomalies, systemic ventricular dysfunction is thought to be the major cause of exertional dyspnea. MRI is a useful non-invasive method for the rapid evaluation of cardiac morphology. (Jpn Heart J 2001; 42: 645-649)

Key words: Corrected transposition of the great arteries, Magnetic resonance imaging, Ventricular dysfunction

CONGENITALLY corrected transposition of the great arteries (CCTGA) is a rare cardiac anomaly characterized by atrioventricular (AV) and ventriculoarterial discordance. However, few patients with this anomaly survive past 50 years of age because of associated congenital intracardiac anomalies such as pulmonary stenosis and ventricular septal defect, or systemic AV valve (anatomical tricuspid) regurgitation and subsequent development of systemic (morphological right) ventricular dysfunction or heart block. In this paper, we describe a 65-year-old woman who had CCTGA with AV valve regurgitation. Magnetic resonance imaging (MRI) was useful for diagnosis.
A 65-year-old Japanese woman was admitted to hospital because of exertional dyspnea. She had been asymptomatic until one month earlier, when progressive exertional dyspnea and generalized fatigue developed. Physical examination revealed that she was 158 cm tall and weighed 61 kg, her blood pressure was 118/64 mmHg, and her pulse rate was regular at 66 beats per minute. She had an accentuated second heart sound at the upper left sternal border. A grade 3/6 holosystolic murmur was heard at the lower left sternal border.

A chest radiograph showed mild cardiomegaly with pulmonary congestion and an egg-shaped ventricle (Figure 1). The liver shadow and stomach bubble were located normally. An electrocardiogram showed abnormal Q waves in the right precordial leads, disappearance of septal q waves in the left precordial leads, and inverted T waves in I, aVL, and V3-6 (Figure 2). Holter electrocardiogram showed no evidence of AV block.

Cardiac catheterization was performed one week after the hospitalization. The intracardiac pressures were within normal limits. The cardiac index was 3.4 l min\(^{-1}\) m\(^{-2}\). Selective coronary arteriography demonstrated a reversed coronary arterial pattern without organic stenosis. Systemic ventriculography showed that the left-sided ventricle with heavy trabeculations was morphologically similar to a normal right ventricle (Figure 3). Furthermore, a grade 2/4 AV valve regurgitation and the absence of ventricular septal defect were observed.

Figure 1. Chest radiograph. The cardiothoracic ratio was 52.5%.
**Figure 2.** Electrocardiograph.

**Figure 3.** Systemic (morphological right) ventriculograph in the right anterior oblique view at end-diastole (A) and end-systole (B). Ejection fraction was 53%.
Transthoracic echocardiography showed diffuse hypokinesis of the left-sided (morphological right) ventricular wall, but was not clear enough to provide useful information. A transesophageal echocardiograph revealed AV and ventriculoarterial discordance with moderate systemic AV valve (anatomical tricuspid) regurgitation. However, we did not detect a ventricular septal defect, atrial septal defect or pulmonary stenosis. Left-sided (morphological right) ventricular chamber had a thick and irregular wall without papillary muscles.

MRI clearly revealed findings indicative of CCTGA (Figure 4); the aorta arose from the left-sided (morphological right) ventricle with heavy trabeculations, and the medially placed pulmonary artery arose from the right-sided (morphological left) ventricle. We diagnosed this patient as CCTGA with systemic AV valve regurgitation. The patient was discharged on medical therapy.

**DISCUSSION**

CCTGA is a rare cardiac anomaly with a poor prognosis. Patients without associated cardiac anomalies are even more rare. The majority of patients (74%) have a ventricular septal defect or pulmonary stenosis (74%), or both cardiac anomalies. They often produce congestive heart failure due to systemic AV valve regurgitation and experience gradual deterioration of the systemic ventric-
ular (morphologic right ventricle) function. Occasionally, patients with CCTGA are asymptomatic and survive until adulthood. Our patient is fortunate since she was asymptomatic until 65 years old.

Concern has been expressed as to whether the morphologic right ventricle can support the systemic circulation over prolonged periods in adulthood. Moreover, it is not clear whether the hypertrophied morphologic right ventricle can be supported by the right coronary artery. In this patient, systemic (morphologic right) ventricular failure was the cause of exertional dyspnea. Adult patients with CCTGA do not have a benign condition, and increasing systemic AV valve regurgitation and complete heart block are known deterioration factors of systemic ventricular function.

In our patient, transthoracic echocardiography could not provide adequate cardiac images because of the rotation of the cardiac apex and the presence of lung tissue. On the other hand, MRI clearly demonstrated findings suggesting CCTGA.

In conclusion, the prognosis of CCTGA without severe associated cardiac anomalies is dependent on the appearance of anatomical tricuspid regurgitation and subsequent development of systemic (morphological right) ventricular dysfunction and advanced heart block. Because this patient has no severe associated cardiac anomalies, systemic ventricular dysfunction is thought to be the major cause of exertional dyspnea. In addition, MRI is a useful non-invasive method for the rapid evaluation of cardiac morphology.

REFERENCES