Myocardial Ischemia Induced by Anomalous Aortic Origin of the Right Coronary Artery in a Patient with Atrial Septal Defect

Fumie MAKI, 1 MD, Tomoaki OHTSUKA, 1 MD, Makoto SUZUKI, 1 MD, Yuji HARA, 1 MD, Yuji SHIGEMATSU, 1 MD, Mareomi HAMADA, 1 MD, Kanji KAWACHI, 2 MD, and Kunio HIWADA, 1 MD

SUMMARY

A 27-year-old woman with atrial septal defect (ASD) and a sensation of squeezing in the anterior chest by effort was admitted to our hospital. In addition to the ASD, the coronary angiogram showed an abnormal anomalous position of the right coronary artery. Exercise thallium (Tl)-201 cardiac scintigram with an electrocardiogram clearly detected myocardial ischemia in the inferior area. In the operative findings, the orifice of the right coronary artery was positioned high above the commissure between the right and left sinuses of Valsalva, and it ran between the aorta and pulmonary trunk. Considering myocardial ischemia possibly caused by the anomalous origin of the right coronary artery, a coronary artery bypass graft (CABG) was simultaneously performed to the right coronary artery with direct closure of ASD. The myocardial ischemic finding in the inferior area disappeared after the operation, and she was also relieved from the chest pain. In view of these findings, we suggest that an active combination treatment such as CABG and ASD closure is highly successful in a patient with a threatening coronary anomaly and congenital heart disease. (Jpn Heart J 2001; 42: 371-376)

Key words: Coronary anomaly, Atrial septal defect, Myocardial ischemia

AN association between the anomalous aortic origin of coronary arteries and fatal cardiac events such as acute myocardial infarction, angina pectoris, syncope, fatal arrhythmias and sudden death has been reported.1-6) However, only one necropsy case of sudden death with a combination of an anomalous origin of the right coronary artery and atrial septal defect (ASD) has been reported.7) We present the first case of an anomalous origin of the right coronary artery combined with ASD whose angina pectoris was relieved after an appropriately placed coronary artery bypass graft (CABG) and direct closure of the ASD.

From 1 the Second Department of Internal Medicine, 2 Second Department of Surgery, Ehime University School of Medicine, Ehime, Japan.
Address for correspondence: Tomoaki Ohtsuka, MD, Second Department of Internal Medicine, Ehime University School of Medicine, Onsen-gun, Ehime, 791-0295, Japan.
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CASE REPORT

A 27-year-old woman suspected of having an ASD was referred to our hospital. She had been suffering from a sensation of squeezing in the anterior chest since 17 years old. It usually occurs on exertion and lasted for a few seconds to 10 minutes. On auscultation, the second sound was split, and unaffected by respiration. The resting electrocardiogram on admission showed an incomplete right bundle branch block. The echocardiogram revealed enlargement of the right ventricle, and a Doppler echocardiogram detected ASD flow. Cardiac catheterization demonstrated a 17 mm ASD. The pulmonary artery pressure was not increased, but the rates of Qp / Qs and Rp / Rs were 1.47 and 0.028, respectively, and the reversed shunt rate was 32.7% as shown in the Table. Coronary angiogram showed a normal left coronary artery, but the orifice of the right coronary artery was above the sinus-tubular junction (Figure 1) and the right coronary artery was slightly squeezed due to compression by the aorta and pulmonary trunk. To elucidate the association of anomalous aortic origin of the right coronary artery with her chest squeezing, an exercise thallium (Tl)-201 cardiac scintigram was performed. As shown in Figure 2A, ischemic redistribution in the inferior area was identified. We considered that the myocardial ischemia was possibly caused by an anomalous aortic origin of the right coronary artery, and that surgery for the anomalous origin of the right coronary artery was needed in addition to closure of the ASD. In the operative findings, the orifice of the right coronary artery was

<table>
<thead>
<tr>
<th>Variables</th>
<th>Value</th>
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<tbody>
<tr>
<td>RA (mean, mmHg)</td>
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</tr>
<tr>
<td>RV (sys/EDP, mmHg)</td>
<td>28/10</td>
</tr>
<tr>
<td>PA (sys/dia/mean, mmHg)</td>
<td>27/11/16</td>
</tr>
<tr>
<td>PAW (mean, mmHg)</td>
<td>12</td>
</tr>
<tr>
<td>Ao (sys/dia/mean, mmHg)</td>
<td>129/83/104</td>
</tr>
<tr>
<td>LV (sys/EDP, mmHg)</td>
<td>117/16</td>
</tr>
<tr>
<td>CI (L/min/m²)</td>
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</tr>
<tr>
<td>Qp/Qs ratio</td>
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</tr>
<tr>
<td>Rp/Rs ratio</td>
<td>0.028</td>
</tr>
<tr>
<td>Shunt ratio (L→R)</td>
<td>0.327</td>
</tr>
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Ao=aortic pressure; CI=cardiac index; EDP=end-diastolic pressure; dia=diastole; LV=left ventricular pressure; PA=pulmonary artery pressure; PAW=pulmonary artery wedge pressure; RA=right atrial pressure; RV=right ventricular pressure; sys=systole.
ANOMALOUS ORIGIN OF RIGHT CORONARY ARTERY

Figure 1. Coronary angiogram. The orifice of the right coronary artery (arrow) positioned high above the sinus-tubular junction.

Figure 2. A: Before the operation, an exercise Tl-201 cardiac scintigram showed ischemic redistribution at the inferior area with ST depression in the II, III, and aVF leads. B: After the operation, the above ischemic findings were not detected. ECG, electrocardiogram; Tl-201, thallium-201 cardiac scintigram.
detected above the commissure between the right and left sinuses of Valsalva, and the right coronary artery was compressed because of the running course between the aorta and pulmonary trunk dilated for the ASD (Figure 3). After direct closure of the ASD, the right internal thoracic artery was bypassed to the right coronary artery. After the operation, an exercise Tl-201 cardiac scintigram was conducted again to assess myocardial ischemia. In striking contrast to the pre-operative finding, the myocardial ischemia had disappeared and she was free from angina pectoris (Figure 2B).

DISCUSSION

This report describes a young woman with combined anomalous origin of the right coronary artery and ASD. Bypass grafting for the right coronary artery concomitant with direct closure of the ASD successfully improved her myocardial ischemia.

Anomalous origin of the coronary artery has been reported as the cause of angina pectoris, arrhythmia, syncope and fatal myocardial infarction.1-6) Its incidence ranges from 0.61% to 1.3%.8,9) Concerning the type of anomalous origin of the coronary artery, sudden death was the most common when the left coronary artery ran between the aorta and pulmonary trunk.10) This type of the anomalous
running was also demonstrated in 25% of the anomaly of the right coronary artery from the left sinus of Valsalva.\textsuperscript{10} These anomalies were sometimes associated with other congenital heart diseases, including bicuspid aortic valve, mitral valve prolapse, and ventricular septal defect.\textsuperscript{8, 11} However, to our knowledge, we have presented here the first case of an anomalous origin of the right coronary artery combined with ASD except for the autopsy case of sudden death reported by Kragel and Roberts.\textsuperscript{7}

In the present case, the orifice of the right coronary artery was highly positioned (so-called high take-off) almost to the commissure between the right and left sinus of Valsalva. The right coronary artery was compressed because of its course running between the aorta and pulmonary trunk. According to Kragel and Roberts,\textsuperscript{7} this is the most common type of anomaly of the right coronary artery, and we considered the following as possible mechanisms for the myocardial ischemia seen in our patient. First, high take-off may be one reason for the myocardial ischemia because the sufficient quantity of blood stored in the cusp cannot be used effectively. Second, compression of the right coronary artery by the aorta and pulmonary trunk may obstruct the coronary blood flow, and thus, induce myocardial ischemia in accordance with coronary squeezing. Third, overload of the right ventricle due to the ASD may involve an abnormality in coronary blood flow of the right coronary artery by enlargement of the right ventricle. Bypass grafting for the right coronary artery concomitant with the direct closure of the ASD relieved her angina pectoris. Exercise TI-201 myocardial scintigrams revealed that surgical repair by CABG and ASD closure clearly cured the myocardial ischemia in our patient. Therefore, simultaneous surgery for the coronary anomaly and congenital heart disease may be the best way to prevent a future fatal cardiac event.

**REFERENCES**