THREE MAIN CORONARY ARTERIES TO PULMONARY ARTERY FISTULA

Kiyoo Mori, M.D., Tamehisa Onoe, M.D., and Takio Ooka, M.D.

A case of a 19-year-old woman with coronary artery fistula was reported. She was asymptomatic throughout her life. Physical examination revealed a continuous murmur along the left sternal border. Echocardiograms and left ventriculograms showed mitral valve prolapse. Selective coronary arteriograms disclosed arteriovenous fistula between branches of the right, the left anterior descending and the left circumflex coronary artery and the main pulmonary trunk. We believe that this is the first case report of an unusual form of anomalous coronary-pulmonary artery communication: three main coronary vessels participate in the fistula.

Congenital coronary artery fistula is an uncommon cardiovascular malformation. In recent years, however, with the introduction of coronary cineangiography the lesion has been diagnosed during life and almost 300 cases have been described since it was originally described by Krause in 1865. As far as we could identify in these reports, no patient with three main coronary vessel involvement has been reported. Here, we would like present a case of anomalous communications between three main coronary arteries and main pulmonary trunk with mitral valve prolapse syndrome.

CASE REPORT

A 19-year-old female was admitted to the Ishikawa Prefectural Central Hospital for evaluation of a cardiac murmur. She has been asymptomatic throughout her life. At the time of admission, physical examination revealed blood pressure of 108/50 mmHg and a pulse rate of 80 beats per minute. The only significant abnormality discovered was a grade II/VI continuous murmur along the left sternal border. This murmur was best heard at the 2nd intercostal space. With methoxamine the continuous murmur increased and lengthened. With amyl nitrite the systolic component of the murmur slightly decreased and the diastolic disappeared (Fig. 1). Chest X-ray film revealed no abnormality. Electrocardiograms recorded at rest and after a Master's two-step test were within normal limit. Echocardiograms suggested mitral valve prolapse (Fig. 2-a). Cardiac catheterization revealed normal pressure and oxygen saturation in all intracardiac chambers and great vessels (Table I). Cardiac index was 4.52 L/min/m². Left ventriculography disclosed posterior mitral valve prolapse without mitral regurgitation (Fig. 2-b, c). The asynergy of the left ventricular wall was not noted. Selective coronary arteriograms disclosed three anomalous vessels originating from the conus branch of the right coronary artery, the left anterior descending coronary artery and the left circumflex coronary artery.
TABLE I CARDIAC CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>Site</th>
<th>Blood Pressure mmHg</th>
<th>O₂ Saturation %</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>(7)</td>
<td>85</td>
</tr>
<tr>
<td>RV</td>
<td>27/1 (14)</td>
<td>89</td>
</tr>
<tr>
<td>main PA</td>
<td>20/7 (14)</td>
<td>80</td>
</tr>
<tr>
<td>left PA</td>
<td>18/11 (13)</td>
<td>80</td>
</tr>
<tr>
<td>right PA</td>
<td>19/8 (13)</td>
<td>83</td>
</tr>
<tr>
<td>PCW</td>
<td>(10)</td>
<td>-</td>
</tr>
<tr>
<td>IVC</td>
<td>-</td>
<td>87</td>
</tr>
<tr>
<td>SVC</td>
<td>-</td>
<td>81</td>
</tr>
<tr>
<td>LV</td>
<td>120/9 (23)</td>
<td>-</td>
</tr>
<tr>
<td>Aorta</td>
<td>120/70 (95)</td>
<td>96</td>
</tr>
</tbody>
</table>

Cardiac index: 4.52 L/min/m²

RA = right atrium, RV = right ventricle, PA = pulmonary artery, PCW = pulmonary capillary wedge, IVC = inferior vena cava, SVC = superior vena cava, LV = left ventricle, ( ) = mean

DISCUSSION

Coronary to pulmonary artery fistulas account for only 20% of all coronary artery fistulas. The lesion is more frequently seen in the right coronary artery than the left and both coronary arterial involvement is rarest. The occurrence is 55%, 35% and 5%, respectively. As far as we could identify in literatures, coronary artery fistulas include communications between only one or two coronary vessels and the pulmonary artery. In the case of both coronary arterial involvement, the abnormal feeding vessels originated from the right coronary artery and one of the following arteries; the left anterior descending, the left circumflex artery or the left main trunk. We believe that this is the first case report of an unusual form of coronary artery fistula: each of three main coronary trees gave rise to anomalous branches draining into the main pulmonary trunk.

Clinically, patients with a coronary artery fistula may be asymptomatic as our case, or present angina pectoris or heart failure. Most of the patients present characteristic continuous murmur located in areas other than the usual location of the continuous murmur of the patent ductus arteriosus. Sakakibara et al. divided coronary artery fistula into five types according to the chamber or vessel into which they drain: into the right atrium, right ventricle, pulmonary

and all were draining into the main pulmonary trunk (Fig. 3).

Surgical treatment was not indicated as the patient had no symptoms, the blood flow through these abnormal vessels was a little and mitral regurgitation due to mitral valve prolapse was none.

Fig. 2. Echocardiogram and left ventriculograms. The M mode echocardiogram suggests mitral valve prolapse (arrows) (a). Selective left ventriculographic frames at systole in the right anterior oblique projection (b) and the left anterior oblique projection (c) demonstrate posterior leaflet prolapse (arrows).

Fig. 3. Selective coronary arteriograms of the left coronary artery in the lateral projection (left) and the right coronary artery in the left anterior oblique projection (right). Anomalous vessels originating from the left anterior descending (A), the left circumflex (B) and the right coronary artery (C), which drain into the pulmonary artery, are shown. LAD = left anterior descending coronary artery, LCX = left circumflex coronary artery, RCA = right coronary artery

artery, left atrium and left ventricle. When the fistula is in communication with the pulmonary artery, the maximal point of the murmur is heard at the second or third left intercostal space. In our case, the maximal point was at the second intercostal space.

Most cases of coronary artery fistulas occur as isolated anomalies. Some occur as a consequence of other cardiac malformation, such as pulmonary or aortic atresia. Crawford reported a case of coronary artery fistula associated with myocardial infarction and he stated that it was acquired secondarily from the coronary obstructions. In the present case, mitral valve prolapse was demonstrated by echocardiography and left ventriculography. Gentzler et al. reported congenital absence of the atroventricular groove branch of the left circumflex artery in a patient with mitral valve prolapse. Others have shown mitral valve prolapse in coronary heart disease. Jeresaty stated that in patients with secondary mitral valve prolapse, clicks were not heard and the echo showed no prolapse. In the present case, phonocardiograms did not demonstrate click, but echocardiograms showed mitral valve prolapse. It remains unknown whether mitral valvular dysfunction was secondary to papillary muscle damage in relation to coronary artery fistula or due to idiopathic myxomatous change of mitral valve.

Acknowledgement

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REFERENCES


