Sudden Deterioration of Aortic Regurgitation due to Rupture of a Raphal Cord on the Conjoined Cusp

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A 57-year-old man was admitted to hospital for acute myocardial infarction associated with mild aortic regurgitation, which was successfully treated by intracoronary thrombolysis. Twenty-four days later, he suffered from another chest pain attack without any electrocardiographic ST-T changes. The coronary angiogram did not show any significant lesions, but the aortic root angiogram showed massive aortic regurgitation. Surgery revealed a bicuspid aortic valve with a conjoined cusp that had a fenestrated raphe torn away from the aortic wall and prolapsing into the left ventricle. The aortic valve was successfully replaced with a St Jude Medical mechanical valve prosthesis. The pathological significance of the intact raphal cord and the rupture remains an unsolved problem. This is the first reported case in which an increase of aortic regurgitation due to a ruptured raphal cord supporting the conjoined cusp was confirmed by a serial root angiogram. (Jpn Circ J 2000; 64: 477-480)

Key Words: Aortic regurgitation; Bicuspid aortic valve; Conjoined cusp; Raphal cord

A bicuspid aortic valve usually shows calcific stenotic lesions in the aged and sometimes regurgitant lesions in younger patients. Infective endocarditis can often affect the bicuspid valve, which results in aortic regurgitation (AR). We present an exceptionally rare form of AR that deteriorated due to a ruptured raphal cord of the conjoined cusp and relevant literature will be reviewed.

Case Report

A 57-year-old man was admitted to hospital complaining mainly of a sudden onset of chest pain. He had a 20-year history of hypertension and hyperlipidemia, but no history of heart disease. His blood pressure was 120/70 mmHg and regular pulse rate was 76 beats/min. His body temperature was 35.9°C. Cardiac auscultation revealed a to-and-fro murmur at the third left sternal border with an intensity of Levine 2/6. Laboratory examination on admission showed a white blood cell count of 8,300 and normal C-reactive protein of 0.30 mg/dl. Creatine kinase and aspartate aminotransferase were 99 IU/L and 221 IU/L, respectively. During the post-admission course, maximum values of these enzymes were 854 IU/L and 1201 IU/L, respectively. The electrocardiogram showed sinus rhythm, left ventricular hypertrophy and elevated ST-T segment in leads of II, III and aVr. Chest roentgenogram showed an increased cardio- thoracic ratio of 54.5%. Emergency coronary angiography showed a localized filling defect in the proximal portion of the left anterior descending artery, which was successfully recalanized after an intracoronary injection of 480,000 U of urokinase. A 50% stenotic lesion was located in the proximal portion of the right coronary artery. Aortic root angiography showed grade II/IV AR (Fig 1A) and left ventriculography showed an increased end-diastolic volume index of 144.8 ml/m², ejection fraction of 83.5%, and hypokinesis of the apical-inferior wall. Echocardiography showed mild AR without evidence of valve prolapse.

The patient recovered without incident, but 24 days later, he again suffered another attack of chest pain and became dyspeptic on exertion. He was readmitted for evaluation of the coronary artery. Laboratory examination showed a white blood cell count of 4,800 and C-reactive protein of 0.30 mg/dl. His body temperature was 36.0°C. His bilateral femoral pulse was bounding and blood pressure measurement showed decreased diastolic blood pressure of 40 mmHg and normal systolic pressure of 130 mmHg. The intensity of the aortic valve regurgitant murmur had increased to Levine 3/6. The left anterior descending artery was not remarkable on coronary angiography, but aortic root angiography showed increased regurgitation of grade III/IV (Fig 1B). Left ventriculography showed an end-diastolic volume index of 146.1 ml/m² and ejection fraction of 78.0%. Pressure studies showed an elevated left ventricular end-diastolic pressure of 13 mmHg. Transthoracic echocardiography showed massive AR with the prolapse of the anterior cusp and its dyscoaptation with the posterior cusp. The abnormally moving structure that is often observed in cases of ruptured cords was not detected in spite of the close examination (Fig 2).

Surgery revealed a bicuspid aortic valve. The fibrous strand that suspended the midportion of the conjoined cusp was torn away from the aortic wall. The conjoined cusp located in the anterior position and the left and right coronary arose from the sinus on both sides of the fenestrated raphe. The aortic valve was replaced with a 25-mm St Jude Medical mechanical valve prosthesis. The ruptured end of the raphal cord did not show any sign of infection such as thrombotic vegetation or plump club-like thickening (Fig 3). Histopathological examination of the torn raphal cord confirmed the absence of endocarditis, but did show

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Fig 1. (A) Aortic root angiogram of the right oblique view on the first admission shows aortic regurgitation of grade I/IV in Sellers' classification (left side). (B) Aortic root angiogram at 24 days after the first admission shows increased aortic regurgitation of grade III/IV (right side).

Fig 2. The echocardiography shows thick aortic valves in which anterior cusp is prolapsed, preventing diastolic coaptation. Vegetation or abnormal strand was not detected.

myxomatous degeneration and calcification (Fig 4). The postoperative course was uneventful and the patient remains well without chest pain.

**Discussion**

The congenital bicuspid aortic valve is characterized by 2 semilunar cusps, which are termed the single and the conjoined cusp, the latter usually wider than the former. In most cases there is a raphe from the aortic wall onto the aortic surface of the conjoined cusp. An exceptionally tall raphe, or raphal cord, extends from the aorta to either the upper free edge or the base of the conjoined cusp and is fenestrated beneath the cord. The raphal cord is usually single, but sometimes multiple cords are present. The incidence of raphal cords in non-infected bicuspid aortic valves showing pure regurgitation is 24%! Walley et al

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reported 17 cases (5.8%) of surgically excised bicuspid aortic valves with stenosis and/or regurgitation that had a fenestrated raphe in the conjoined cusp. A similar anatomic arrangement was observed in the pulmonary valve of a patient with tetralogy of Fallot.

Hemodynamic deterioration of a normally functioning congenital bicuspid valve increases with age. Calcification with stenosis occurs in nearly all patients after the age of 60. Aortic regurgitation in younger adults may be more evident than calcific stenosis and usually results from diastolic prolapse of a wider conjoined cusp. Infective endocarditis, which usually results in a regurgitant lesion, is a recognized potential complication of a congenital bicuspid aortic valve. Another cause of AR is aortic root dilatation, because the bicuspid valve is less able to tolerate an increase in the aortic root diameter than the tricuspid valve. Proximal aortic dissection, for which a bicuspid aortic valve is a well-recognized risk factor, is also an important cause of acute AR. More exceptionally, rupture of the fenestrated raphe, especially if it is attached at the free edge of the cusp, can lead to acute and massive AR resulting in left heart failure. In that case, the conjoined cusp has usually been affected by chronic AR and the raphal cord supporting the wider conjoined cusp has been exposed to greater mechanical stress during the diastolic phase because of valve prolapse. To our knowledge, 6 such cases have been reported (Table 1). Cater et al reported a 59-year-old man with AR whose autopsy showed a ruptured raphal cord allowing the conjoined cusp to prolapse. The case reported by Becker et al involved a 45-year-old man with mitral and aortic regurgitation. The aortic lesion was chronic and the mitral lesion was speculated to be secondary to left ventricular dilatation in the presence of AR. The aortic conjoined cusp prolapsed into the left ventricular cavity because the raphal cord supporting the valve ruptured. Histopathological examination showed

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**Table 1 Reported Cases of Raphal Cord Rupture**

<table>
<thead>
<tr>
<th>Case no. (ref. no.)</th>
<th>Age/Sex</th>
<th>Symptom</th>
<th>AR</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (9)</td>
<td>59/M</td>
<td>NS</td>
<td>NS</td>
<td></td>
<td>Autopsy</td>
</tr>
<tr>
<td>2 (10)</td>
<td>45/M</td>
<td>Dysnea</td>
<td>3/3</td>
<td>AVR+MVR</td>
<td>Alive</td>
</tr>
<tr>
<td>3 (1)</td>
<td>NS</td>
<td>NS</td>
<td>Acute</td>
<td></td>
<td>NR</td>
</tr>
<tr>
<td>4 (11)</td>
<td>53/M</td>
<td>Chest discomfort</td>
<td>3/4</td>
<td>AVR</td>
<td>Alive</td>
</tr>
<tr>
<td>5 (2)</td>
<td>53/M</td>
<td>Dysnea, angina</td>
<td>4/4</td>
<td>AVR</td>
<td>NS</td>
</tr>
<tr>
<td>6 (2)</td>
<td>58/M</td>
<td>Dysnea</td>
<td>4/4</td>
<td>AVR</td>
<td>NS</td>
</tr>
<tr>
<td>7 (present case)</td>
<td>57/M</td>
<td>Dysnea, angina</td>
<td>3/4</td>
<td>AVR</td>
<td>Alive</td>
</tr>
</tbody>
</table>

AR: aortic regurgitation; AVR: aortic valve replacement; MVR: mitral valve replacement; NS, not stated.

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acumulation of mucopolysaccharides, but no sign of infection. Hamada et al reported a 53-year-old man who suddenly developed AR due to a ruptured raphal cord of a conjoined cusp, which was successfully replaced. Olson reported one case of a ruptured raphal cord of the conjoined cusp in 54 cases of bicuspid aortic valve with AR, although the details were obscure! Walley reported 2 cases of bicuspid aortic valve with ruptured raphal cord in a retrospective pathological study of surgically excised bicuspid valves. The cause of AR in 1 case was suspected to be raphal cord rupture because of the macroscopic findings at the end of the cord, and the other case had a fractured raphal cord with severe calcification. Both cases had had massive AR and their valves had been replaced.

In the present case, rupture of the raphal cord is considered to be the cause of the sudden increase in AR. The initial acute coronary event and coronary thrombosis therapy are probably unrelated to the rupture of the raphal cord. Increased AR can result in abnormal stress on the raphal cord, but in the present case the hemodynamic deterioration resulted from an acute myocardial infarction due to an elevated left ventricular end-diastolic pressure, which decreases the diastolic pressure gradient between the aorta and left ventricle and decreases regurgitant blood flow. Trauma by the catheter tip was also unlikely, because intraprocedural hemodynamic deterioration was not detected during the continuous monitoring, and there was no suspicion of postprocedural infective endocarditis, which is a rare complication of cardiac catheterization. In addition to the preexisting hypertension in the present patient, fibrosis, calcification and myxomatous degeneration of the raphal cord, which is the first to be affected by such degenerative changes, are the most likely causative factors for acute disruption of the raphal cord.

The mean age of the reported cases with raphal cord rupture was 54.2 years old. Aortic regurgitation due to the raphal cord rupture seldom occurs in the younger generation without degenerative changes of the cord. Previously reported cases with ruptured raphal cords had long histories of mild or moderate AR and dilated aortic annulus, but it was not proven that the increase in AR was because of the ruptured raphal cord. The initial regurgitation may have resulted from impaired cusp coaptation due to the aortic annular dilatation. Although an intact raphal cord tends to prevent prolapse of the conjoined cusp, some reports show that it could disturb the coaptation between the conjoined cusp and the single cusp, which prolapses into the left ventricle due to aortic annular dilatation and subsequently AR occurs. The raphal cord rupture was not diagnosed preoperatively in any of the previously reported cases including the present case. An abnormally moving fibrous band in the left ventricle is thought to be highly suggestive of raphal cord rupture in the absence of signs of infected endocarditis, and may be detected with higher sensitivity by transeosophageal echocardiography than by the conventional transthoracic approach.

We conclude that rupture of the raphal cord of the conjoined cusp in a bicuspid aortic valve can be a cause of sudden deterioration of AR, similar to the rupture of the fibrous strands in fenestrated tricuspid aortic valves. The massive regurgitation due to a ruptured raphal cord is prone to induce rapidly progressive hemodynamic deterioration, so earlier aortic valve replacement is indicated.

References