Aortic Regurgitation Due to Fibrous Strand Rupture in the Fenestrated Left Coronary Cusp of the Tricuspid Aortic Valve

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SUMMARY

Fenestration-related massive aortic regurgitation is rare. The underlying mechanism is reported to be rupture of the fenestrated fibrous strand, and most ruptured cords have been reported in the bicuspid valve or in the right coronary cusp of the tricuspid aortic valve. We encountered a rare case of acute aortic regurgitation due to fibrous strand rupture in the fenestrated left coronary cusp. Preoperative echocardiography detected left coronary cusp prolapse, and operative findings revealed rupture of a fibrous strand in the left coronary cusp. For cases such as this, preoperative echocardiography would be useful for appropriate diagnosis. (Int Heart J 2014; 55: 550-551)

Key words: Aortic valve regurgitation

Fenestration in the aortic valve is not uncommon and is often observed in normal subjects. In this type of aortic valve, the fibrous strand, which is said to be an embryonic remnant during semilunar cusp formation, is a supportive tissue that maintains aortic valve coaptation. Fibrous strand rupture causes acute aortic regurgitation (AR) and has been reported to occur in the congenital bicuspid valve or in the right coronary cusp of the tricuspid valves.1 Here we report the case of a patient with a fenestrated tricuspid aortic valve with a fibrous strand. The patient had a history of hypertension and suffered from acute AR due to fibrous strand rupture in the left coronary cusp.

CASE REPORT

A 76-year-old man underwent aortic valve replacement with a bioprosthetic valve (23 mm) for acute AR. He had a history of hypertension, and his blood pressure was poorly controlled despite medication. Chest X-rays revealed gradual worsening of cardiomegaly since the initiation of medication therapy at 66 years of age. He had no history of rheumatic heart valve disease.

At the age of 75 years, moderate AR was identified with echocardiography. However, he did not have any definite symptoms at that time, and medication therapy was continued instead of surgical treatment. Dyspnea on exertion became gradually more severe, and he was transferred to our hospital due to acute heart failure. His blood pressure was 220/96 mmHg and the control of blood pressure was very poor. His body temperature was 36.7°C, and no inflammatory response was observed. Auscultation revealed a diastolic murmur (Levine III/VI) at the right second intercostal sternal border. Chest X-rays revealed cardiomegaly (cardiothoracic ratio = 65%) and pulmonary edema. His electrocardiogram showed a strain T pattern (concentric hypertrophy) in V5 and V6. Serum brain natriuretic peptide (BNP) was extremely high at 426 pg/mL. Echocardiography showed a dilated left ventricular chamber (left ventricular diastolic dimension: 57 mm) with fair contractile function (ejection fraction = 50%). The aortic valve was tricuspid; however, fenestrated cusps with fibrous strands were detected in the right coronary cusp and noncoronary cusp. The left coronary cusp was prolapsed and had dropped into the left ventricular chamber. A ruptured fibrous strand was attached to the left coronary cusp (Figure 1). Aortic annular size was 23 mm by echocardiography, and regurgitation jet which reached to the papillary muscle was observed in the left coronary cusp. Pressure half time was 163 ms, indicating severe regurgitation. Pulmonary artery pressure measured by catheter examination was 50/24 (37) mmHg, and pulmonary capillary wedge pressure (PCWP) was 20 mmHg. Aortography showed severe AR (Seller’s grade IV), and coronary angiography revealed intact coronary arteries.

The patient underwent aortic valve replacement for severe acute AR. During the operation, fibrous strands were found in all cusps (Figure 2). The right and noncoronary cusps formed fenestrations, and the left coronary cusp had a ruptured fibrous strand (Figure 2). We chose to use a bioprosthetic valve (23 mm) for aortic valve replacement.

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Aortic regurgitation (AR) caused by fibrous strand rupture is rare. This problem was first reported in the literature in 1981. It was thought to be associated with bicuspid aortic valves and rarely with tricuspid aortic valves; however, several reports have recently documented tricuspid aortic valve cases. Most previous reports described ruptured fibrous strands in the right or right and noncoronary cusps, although patterns of fenestration or fibrous strand attachment differed in each case. These reports have mostly been from Japan and other Asian countries suggesting that some form of heredity involvement may exist with regard to fibrous strand formation.

In the present case, fibrous strands supported the aortic valve and maintained coaptation of the valve (Figure 3). Poorly controlled hypertension may have led to mechanical stress on the fibrous strands, and continuous tension to this supportive tissue may have gradually weakened the strand, resulting in AR due to cord rupture. This type of support tissue has been referred to as a “fibrous strand,” “fibrous band,” or “retention chord” in the literature, with no definitive name. Fibrous strands typically appear with fenestrations and may occur in normal healthy subjects; however, most of them do not suffer from AR unless the fibrous strand ruptures. In the present case, in addition to the ruptured strand in the left coronary cusp, the right and noncoronary cusps maintained fibrous strands and valve coaptation, forming fenestrations (Figure 3).

The essential anatomic features of aortic valve disease can be clearly defined by high-resolution two-dimensional echocardiography because ruptured fibrous strands are highly echogenic objects attached to valve leaflets. They may also be observed as floating objects located between a valve leaflet and the aortic wall. Furthermore, an enlarged aortic annulus > 30 mm in diameter is often observed with echocardiography. Transesophageal echocardiography and transthoracic echocardiography are reported to be useful for the detection of fibrous strand rupture. In the absence of any other obvious cause, physicians must consider the possibility of a fenestrated aortic valve in patients with chronic AR or sudden deterioration of regurgitation.

We encountered a case of acute AR as the result of fibrous strand rupture, which we were able to successfully diagnose prior to surgery with echocardiography. If Asian patients do not have a history of rheumatic heart valve disease but develop acute AR and preoperative echocardiography shows AR with a fibrous strand near the valve leaflet, fibrous strand rupture should be considered.

### References