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Bentall Operation in a Patient with a Unicommissural Unicuspid Aortic Valve

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A unicuspid aortic valve is a rare congenital malformation that frequently presents with valvular dysfunction and dilatation or aortic aneurysm, requiring combined aortic valve surgery and aortic repair. Some patients show severe valve calcification extending into the interventricular septum, possibly resulting in damage to the conduction system during debridement for valve replacement. We present a rare case of severe aortic stenosis with a unicommissural unicuspid aortic valve diagnosed by preoperative transesophageal echocardiography in a 36-year-old man. After composite graft replacement of the aortic valve, aortic root, and ascending aorta, a permanent pacemaker was placed because of postoperative complete heart block.

Key words: 1. Aortic valve stenosis
2. Aortic aneurysm
3. Congenital heart defects
4. Heart block
5. Pacemaker

Case report

A 36-year-old man was admitted because of dyspnea and chest pain for 1 month prior to presentation. A grade 4/6 systolic murmur was auscultated at the right second intercostal space. Initial chest roentgenography showed mediastinal widening, and electrocardiography (ECG) showed a 2:1 atrioventricular block (Fig. 1A). Transthoracic echocardiography (TTE) showed normal left ventricular systolic function (ejection fraction, 62%) with concentric left ventricular hypertrophy and typical findings of severe aortic stenosis, including mild aortic regurgitation, extensive calcification of the aortic valve, a mean transaortic valve pressure gradient of 49 mm Hg, and an aortic valve area of 0.8 cm^2. However, there was no definite commissure or raphe between the aortic cups, implying a unicuspid aortic valve (UAV). Subsequent transesophageal echocardiography (TEE) revealed a UAV with a single posterior commissure (Fig. 1B–E). Computed tomography showed aneurysms of the aortic root (5.0 cm in diameter) and ascending aorta (5.5 cm in diameter) (Fig. 2).

An incision made through the ascending aorta revealed an extensively calcified aortic valve (Fig. 3A). The aortic valve was morphologically unicuspid with a single, unfused commissure that had a slit-shaped appearance, and it was distinguishable from a bicus-

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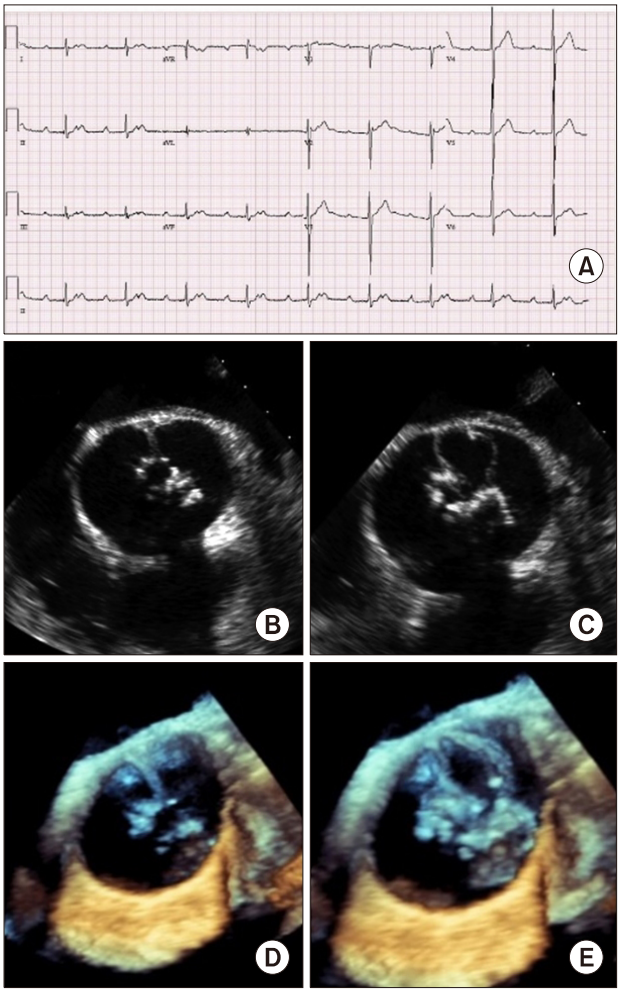

Fig. 1. (A) Electrocardiography performed at admission shows a 2:1 atrioventricular block. (B-E) Preoperative transesophageal echocardiography shows a unicuspid aortic valve with a single posterior commissure.

pid aortic valve (BAV) (Fig. 3B). The diseased aortic valve was carefully removed using a surgical blade and rongeur. However, extensive aortic annular calcification had deeply invaded the annulus, especially on the left coronary cusp, and after removal of the calcified tissues, the annular tissue defects were reconstructed with bovine pericardium (Fig. 3C). After removal of the aneurysmal aortic root and ascending aorta, a Bentall operation was performed using a 27-mm mechanical valved conduit.

After surgery, ECG showed intermittent complete atrioventricular block, and we considered a temporary pacemaker with an epicardial pacing wire. The patient was weaned from mechanical ventilation on postoperative day 1 and transferred to the general ward on postoperative day 2. He underwent 24-hour continuous Holter monitoring. The minimum heart rate was 44 bpm and the maximum R-R interval was 1.48 seconds. He was discharged on postoperative day 12 without dizziness or fainting. However, he was readmitted because of syncope on postoperative day 21. Intermittent complete atrioventricular block was noted, with a minimum heart rate of 19 bpm and a maximum R-R interval of 14.16 seconds. Thus, a permanent pacemaker was placed.

After discharge from the hospital in a stable condition, the patient was placed under appropriate oral anticoagulation therapy and was followed up at an outpatient clinic. Follow-up echocardiography performed 2 years after surgery revealed a diminished left ventricular mass index (from 121.00 to 77.14 g/m^2).

The Institutional Review Board of Sanggye Paik Hospital approved the present case report (IRB approval no., SGPAIK 2019-02-016), and informed consent was obtained from the patient.

Discussion

A UAV, which occurs about 50-fold less often than a BAV, was first reported by Edwards [1] in 1958. Although a UAV is the second most frequent aortic valve anomaly, it is an extremely rare congenital malformation, with an incidence of roughly 0.02% among patients referred for echocardiography [2].

The weight of a UAV is greater than that of a BAV, and extension of calcification into the interventricular septum results in damage to the conduction system during debridement for aortic valve replacement (AVR) [3].

The following 2 UAV subtypes have been described in the literature: pinhole-shaped acommissural UAV and slit-shaped unicommissural UAV [2]. Unicommissural UAV appears to have a less aggressive course than pinhole-shaped acommissural UAV, which is usually accompanied by severe stenosis, with a tendency for symptoms to be present in childhood. In the present case, gross examination of the surgically excised valve showed typical unicommissural valve morphology with extensive calcification.

The preoperative diagnosis of a UAV is rare. In a previous review, only 10% of UAVs were diagnosed using TTE, but TEE might help to identify UAVs [3].

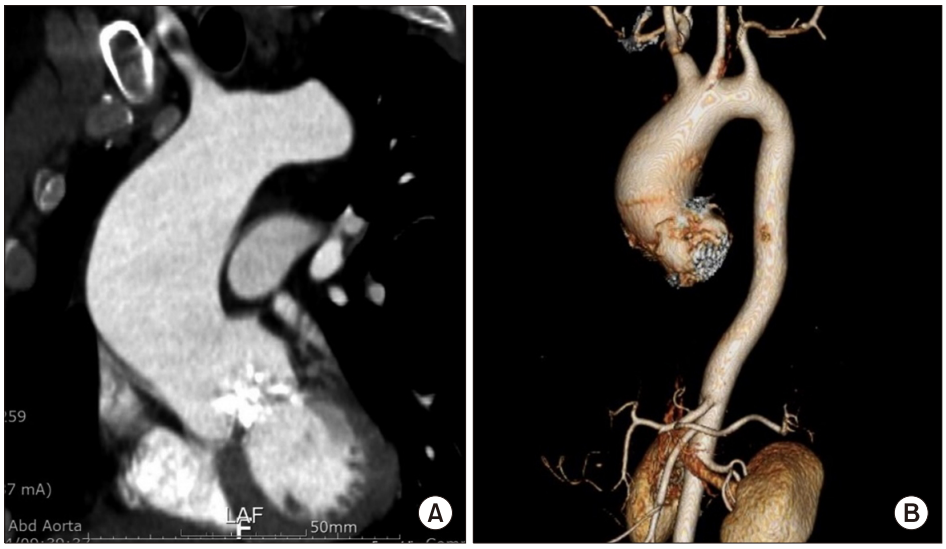

Fig. 2. (A, B) Computed tomography shows aneurysms of the aortic root (5.0 cm in diameter) and ascending aorta (5.5 cm in diameter).

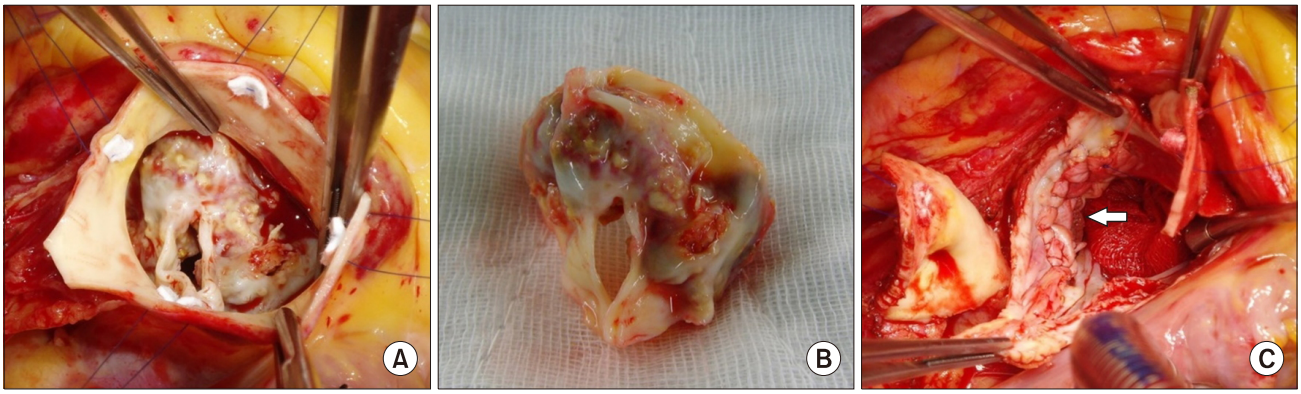

Fig. 3. (A, B) The aortic valve was morphologically unicuspid with a single, unfused commissure that appeared slit-shaped. (C) After removal of calcified tissues, the annular tissue defects were reconstructed with bovine pericardium (arrow).

In the present case, the initial TTE findings indicated a UAV, and additional 3-dimensional TEE confirmed the presence of a typical unicommissural aortic valve. The precise preoperative diagnosis of a UAV can help determine the scope and method of surgery by enabling additional diagnostic procedures.

Although ascending aortic dilatation is frequent in patients with a BAV, some studies have investigated UAV patients and noted ascending aortic dilatation with mild histologic changes of the media [2,3].

In a previous review, 14% of UAV patients showed aortic dilatation or aneurysms and 23% required concomitant aortic surgery at the time of AVR [3]. Aortic annular dilatation is another common feature of a UAV, independent of the diameter of the ascending aorta, suggesting that aortic annular dilatation and ascending aorta aneurysm development are likely due to different pathologic processes. A recent study reported that UAV patients demonstrated thoracic aortic dilatation from the aortic annulus to the proximal arch. Additionally, the authors performed aortic valve surgery combined with aortic repair in 61% of UAV patients and found that long-term survival was better in these patients than in those who underwent valve surgery alone [4].

In the present case, the ascending aortic diameter was 55 mm; that of the aortic annulus was 36 mm, and that of the sinus portion was 50 mm. Considering the patient's age, we decided that composite root replacement would be better than AVR

alone or AVR with ascending aortic replacement.

The valve repair procedure is an important issue in aortic valve disease. Few cases of valve repair in patients with a UAV have been reported [5,6]. Most UAV patients showed severe valve calcification, and valve repair should be performed in patients with less calcification and a more favorable morphology for repair.

Another important clinical implication of an extensively calcified UAV is the extension of calcification into the interventricular septum, possibly resulting in damage to the conduction system during debridement for AVR [7]. The mean weight of UAVs (4.36 g) has been reported to be significantly higher than that of BAVs (3.34 g) and tricuspid aortic valves (2.04 g) [8]. Moreover, a greater weight has been shown to be associated with more severe stenosis. Calcification debridement in preparation for valve implantation was undoubtedly the mechanism of injury; hence, caution should be exercised during subvalvular debridement. In the present case, although annular reconstruction was performed at the left coronary sinus area, which was irrelevant to the conduction tissue, the patient eventually needed a permanent pacemaker. The patient's initial ECG showed conduction problems, indicating that extensive calcification might have already damaged the conduction tissues preoperatively.

In conclusion, UAVs are a rare clinical condition that frequently present with ascending aneurysms, requiring combined aortic valve surgery and aortic repair. Surgeons should keep in mind that the extent of calcium deposits might be more severe for UAVs than for BAVs, thereby increasing the risk of conduction tissue injury during surgery.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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