An Adult Case of Unicommissural Unicuspid Aortic Valve Diagnosed Based on the Intraoperative Findings

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Abstract

We herein report an adult case of unicommissural unicuspid aortic valve (UA V). A 59-year-old man, who was noted to have a cardiac murmur at 31 years of age, was admitted to our hospital due to acute heart failure. Severe calcification in the aortic valve with severe low-flow/low-gradient aortic stenosis and moderate aortic regurgitation was observed and thought to be the cause of heart failure, however, the etiology of aortic valve dysfunction was not clear. Aortic valve replacement was subsequently performed, and unicommissural UA V was diagnosed according to the intraoperative findings. UA V is very rare congenital aortic valve disease which is rarely diagnosed preoperatively.

Key words: unicommissural unicuspid aortic valve, low-flow/low-gradient aortic stenosis, aortic valve replacement

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Introduction

Unicuspid aortic valve (UA V) is a very rare congenital disease, with an incidence of 0.02% (1, 2). This condition is divided into acommissural and unicommissural types; the former exhibits severe valve dysfunction and requires treatment from the neonatal period through childhood (3, 4). On the other hand, the unicommissural type often remains asymptomatic until adulthood. UA V is difficult to diagnose preoperatively (5) and is often associated with abnormalities of the aorta and/or cardiac malformations (2, 6-12). Compared with the bicuspid aortic valve (BA V), UA V is considered to progress more rapidly; therefore, a careful follow-up is necessary (5, 10, 13).

We herein report a case of UA V that was preoperatively unidentifiable, but finally successfully diagnosed according to surgical findings.

Case Report

A 59-year-old man was admitted to our hospital with acute heart failure.

He was noted to have a cardiac murmur at a medical checkup at 31 years of age, however, he did not undergo a detailed examination at that time.

During an admission to another hospital for acute heart failure treatment at 56 years of age, a low cardiac function (left ventricular ejection fraction: LVEF 25%, LV diastolic diameter/systolic diameter: LVDd/Ds 67/59 mm), aortic stenosis and regurgitation (ASR) and atrial fibrillation (AF) were observed. Coronary angiography (CAG) revealed no significant stenosis. Surgery was recommended, but he refused at that time. He remained relatively stable at New York Heart Association (NYHA) class II with pharmacotherapy.

The patient was diagnosed with type 2 diabetes at 58 years of age and treated with diet modification and pharmacotherapy.

Three months prior to the admission to our hospital, the patient noticed shortness of breath during his daily activities. Approximately 1 week prior to admission, he suffered from an upper respiratory tract infection, which worsened the symptom and he began to experience shortness of breath

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Upon consultation with a local physician, a chest radiograph revealed cardiac enlargement with pleural effusion and pulmonary congestion. The patient was then referred to our hospital and was admitted for a detailed examination and treatment.

On admission, the patient’s blood pressure was 124/80 mmHg, pulse rate was 114 bpm (irregular), and SpO₂ was 96% with room air. On auscultation, accentuated S₂ and S₃ heart sounds and Levine 4/6 systolic and diastolic murmurs in the 2RSB-3LSB region were noted. There was an increase in the area of cardiac dullness with lateral and downward displacement of the apex beat. Coarse crackles were heard in the bilateral lung fields. Laboratory findings revealed a markedly elevated brain natriuretic peptide (BNP) level of 1,921 pg/mL. A chest radiograph showed cardiac enlargement with bilateral pleural effusion and interstitial pulmonary edema. A 12-lead electrocardiogram revealed AF tachycardia with a heart rate of 130 bpm, and ST segment depression in leads II, III, aVF, V₅, and V₆. Computed tomography (CT) showed severe calcification of the aortic valve, whereas there was no calcification in the thoracic to abdominal aorta. Additionally, we found no aortic abnormalities such as enlargement, aneurysm, or coarctation (Fig. 1). No significant stenosis or abnormalities were observed in CAG (Fig. 2).

Transthoracic echocardiography (TTE) revealed dilatation of the left ventricle (LVDd/Ds 69/64 mm), diffusely decreased left ventricular wall motion, with an LVEF of 17%, and severe calcification in the aortic valve (Fig. 3A and B). The AR jet width, vena contracta, pressure half time and angiography grade were 33%, 0.46 mm, 423 msec and 2 to 3, respectively (Fig. 3C and D) and the pulse pressure was not wide. The aortic max velocity was 3.03 m/sec, and planimetry of the aortic valve area (AVA) was 0.69 cm² (Fig. 4A).

Although stress echocardiography was not performed because of non-sustained ventricular tachycardia, which occurred frequently at that time, on the basis of marked left ventricular (LV) dysfunction with an LVEF of 17% and AVA of 0.69 cm², the stage of AS was considered to be severe LF/LGAS.

The ascending aorta and aortic root were not dilated, and there was no calcification in the thoracic to abdominal aorta (Fig. 1). The physical findings, inflammatory findings, and imaging findings did not support aortitis syndrome. Addi-

Figure 3. Transthoracic echocardiography revealed dilatation of the left ventricle (LVDd / Ds 69/64 mm), diffusely decreased left ventricular wall motion, with an LV ejection fraction of 17% (A, B), and AR jet width, vena contracta, pressure half time and angiography grade of 33%, 0.46 mm, 423 msec and 2 to 3, respectively (C, D). A: Systolic phase, B, C, D: Diastolic phase, LVDd: left ventricular diastolic diameter, LVDs: left ventricular systolic diameter

Additionally, the results of a syphilis test were negative. There were no intraoral aphthous ulcers, skin findings, or eye symptoms characteristic of Behcet’s disease. According to these results, we considered that dilatation of aorta, aortitis syndrome, Behcet’s disease, and syphilis-induced vasculitis were unlikely as the cause of the AR. No ventricular septal defect (VSD) was observed.

Although severe calcification around all commissures caused difficulty in assessing the aortic valve, it appeared that the left lateral (LL: left coronary/right coronary) and posterior (P: left coronary/non coronary) commissures did not open and only the anterior (right coronary/non coronary) commissure opened (Fig. 4B and C). Regurgitation was primarily observed from the center of the aortic valve (Fig. 4D).

Although we could not ascertain the etiology of aortic valve dysfunction, the response to medical treatment for severe LF/LGAS and moderate AR, and acute decompensated heart failure was poor. AVR was subsequently performed using a 27 mm Carpentier-Edwards PERIMOUNT aortic valve (Edwards Lifesciences). The findings of intraoperative transesophageal echocardiography (TEE) were similar to those of TTE (Fig. 4E and F). Additionally, the intraoperative findings (Fig. 5) showed that the LL and P commissures were lower than the right coronary ostium and anterior commissure was of normal height. According to these findings, this case was diagnosed as UAV.

There were no apparent complications during the postoperative period. On postoperative day 44, the LVEF markedly improved to 33% and the BNP level decreased to 149.9 pg/mL. The patient’s status improved to NYHA class II.

Discussion

UAV is a congenital heart disease that was first reported in 1958 by Edwards et al. (14). Compared with BAV, which has a prevalence of 0.9-1.36%, UAV is extremely rare with a prevalence of 0.02% (1, 2). While genetic factors have been considered with BAV (3, 15), no clearly identified genetic abnormalities have been identified for UAV (3), although it is more common among men, with a male to female ratio of 4:1.

UAV is classified into two types, pinhole-shaped acommissural UAV and slit-shaped unicommissural UAV (2). Acommissural UAV presents with severe AS from the early stage (4); it is often associated with symptoms of left heart failure (5) and often requires invasive treatment from the neonatal period (4, 16). On the other hand, unicommissural UAV, which has one commissure with two rudimentary commissures with a broader valvular area, as in our patient, than that in acommissural UAV, is often asymptomatic until 30-50 years of age (17, 18).
It is generally recognized that AS is common in UAV and primarily occurs earlier than AR (2, 5). We considered that our patient initially had severe AS, which progressed to severe LF/LGAS with a gradually worsened LV function and valve cusp degeneration, which then led to the appearance of AR. Accordingly, we believe that the clinical course of
In the present case, a congenital abnormality of the aortic valve was compatible with the finding of a heart murmur in his youth, the lack of calcified lesions in the major vessels despite the observation of severe calcification in the aortic valve, and no other risk factors for arteriosclerosis other than a short duration of type 2 diabetes diagnosed one year previously.

We believed that if there is a possibility of vasculitis due to AR, such as aortitis syndrome, Behcet’s disease, and syphilis-induced vasculitis, then it is necessary to perform contrast-enhanced CT, gallium scintigraphy, and/or positron emission tomography-CT.

In the present case, the preoperative examinations revealed no other concurrent cardiac, aortic or coronary artery abnormalities. Thus, operation was indicated with the diagnosis of symptomatic severe LF/LGAS and moderate AR associated with heart failure of stage D and severe LV dysfunction (19).

The intraoperative findings revealed that the LL and P commissures were below the right coronary artery ostium, and the anterior commissure was of normal height. According to the criteria of Andersen (20), the LL and P commissures were defined as rudimentary. Therefore, this case was ultimately diagnosed as unicommissural UAV.

From our experience in the present case, we propose that the following three points regarding UAV should be clinically noted.

First, the preoperative diagnosis is often difficult. Valvular disease is generally diagnosed by TTE or TEE (18, 21). However, 60% of UAV cases are diagnosed upon inspection of the surgical specimen or an autopsy by establishing the fact that one commissure is of normal height while the other commissures are below the level of the right coronary ostium, i.e., rudimentary by definition (20). The preoperative diagnosis rate of UAV using TTE and/or TEE was reported to be very low (approximately 15%) (2). One potential explanation for the low preoperative diagnosis rate is that fact that the unicommissural UAV features on echocardiography are not widely recognized. Another reason may be that UAV presents with severe calcification, which likely impedes precise imaging. It has been reported that the mean specimen weight is 2.04 g for the tricuspid aortic valve (TAV), 3.34 g for BAV, and 4.36 g for UAV, which is associated with the degree of calcification of the valve (2, 22). Other diagnostic methods, such as CT and magnetic resonance imaging (MRI), were reported to be effective (17, 23, 24). Performing such examinations prior to surgery may have been useful for our patient. Although UAV is rare at 0.02% (1, 2) in the general population, it is observed in 4-5% of patients undergoing surgery for AS (17), and thus, it may not necessarily be rare in these patients. If TTE or TEE was performed upon the suspicion of UAV, then a preoperative diagnosis could have been possible. Thus, we recommend that UAV should be considered as a differential diagnosis for the cause of aortic valve dysfunction.

Second, concurrent abnormalities should be noted. Similar to BAV (25, 26), UAV patients have been reported to have concurrent aortic disease, such as aneurysm in 14%, aortic dissection in 2%, and coarctation of the aorta in 0.4% of cases (2, 6-11). Moreover, concurrent abnormalities of the coronary artery and VSD were reported to occur in 0.8% of patients, and some patients presented with several of these abnormalities (7, 12). Compared with a normal aortic valve, UAV was reported to have a 5-9 times higher risk for aortic dissection (27, 28); it is important to note that this has been reported to occur earlier in UAV than in BAV and TAV (29). Although these complications were not observed in the present patient, systemic preoperative examinations should include an evaluation of the presence of these concurrent abnormalities.

Third, the progression of valvular dysfunction is more rapid than with BAV and TAV. With UAV, AVR is required 10 to 20 years earlier than with BAV and 20 to 30 years earlier than with TAV (5, 10, 13). Therefore, cases with UAV require careful observation.

In conclusion, UAV is a rapidly progressive disease that is often difficult to diagnose preoperatively. Moreover, a careful search for complications other than valvular disease is necessary. When an aortic valve dysfunction is observed, the possibility of UAV should be considered, although it is an extremely rare congenital abnormality.

The authors state that they have no Conflict of Interest (COI).

References

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