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Case Report

Successful Repair of the Quadricuspid Aortic Valve

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Quadricuspid aortic valve (QAV) is rare. Aortic valve replacement is the most frequent treatment for those patients with severe regurgitation or stenosis of the QAV. However, as our comprehension of the aortic valve grows, to repair a dysfunctional QAV becomes more and more feasible. We present a case of successful repair of the QAV using the surgical strategy of bicuspidization.

Keywords: quadricuspid aortic valve, repair

Introduction

Quadricuspid is extremely rare compared with other morphological abnormalities of the aortic valve such as bicuspid or unicuspid. In recent years, many surgeons are struggling to preserve the aortic valve. We share a case of quadricuspid aortic valve (QAV) associated with severe aortic regurgitation and ascending aortic dilatation. Repair of the aortic valve and replacement of the ascending aorta were successfully performed.

Case Report

A 53-year-old man was admitted to our institute during a routine medical examination. Mild hypertension and a transient cerebral ischemic event were noted in his history. Physical examinations revealed a diastolic murmur. The transthoracic echocardiogram (TTE) showed severe aortic regurgitation due to malcoaptation of the leaflets combined with mild aortic stenosis, peak velocity of 2.64 m/s and mean gradient of 16.7 mmHg. The left ventricle end systolic diameter was measured as 4.22 mm, while the ejection fraction was 52.3%. The parameters of the aortic root and ascending aorta were measured on the preoperative CT scan: the aortic annulus was 26 mm, the aortic root was 32 mm, and the ascending aorta was 50 mm. Therefore, the surgery was scheduled later on.

In the operating room, a relative smaller right coronary cusp and an accessory cusp between the left and right coronary cusps were revealed by transesophageal echocardiogram (TEE), and the orientation of the commissures was almost symmetric (**Fig. 1**). The cannulation was performed by the aortic arch (aortic) using the Seldinger technique and the right atrium (two-stage venous). The heart was then arrested by del Nido cardioplegia. After the ascending aorta was transected, the relationship of the leaflets was carefully analyzed: the commissures between the left accessory cusps and the right non cusp commissure were fused by calcified focus; the commissures between the accessory right cups and the left non cusps were retracted respectively by false chordae to the aortic wall. Some small lesions of calcification can be seen on the accessory right commissure. The geometric height of the four cusps was measured to determine the possibility to preserve the valve. Fortunately, all the cusps were larger than 18 mm. The aortic annulus was measured as 23 mm. The aortic root was then dissected carefully to the basal aortic ring; a subcoronary tunnel was created for passing the Dacron ring as

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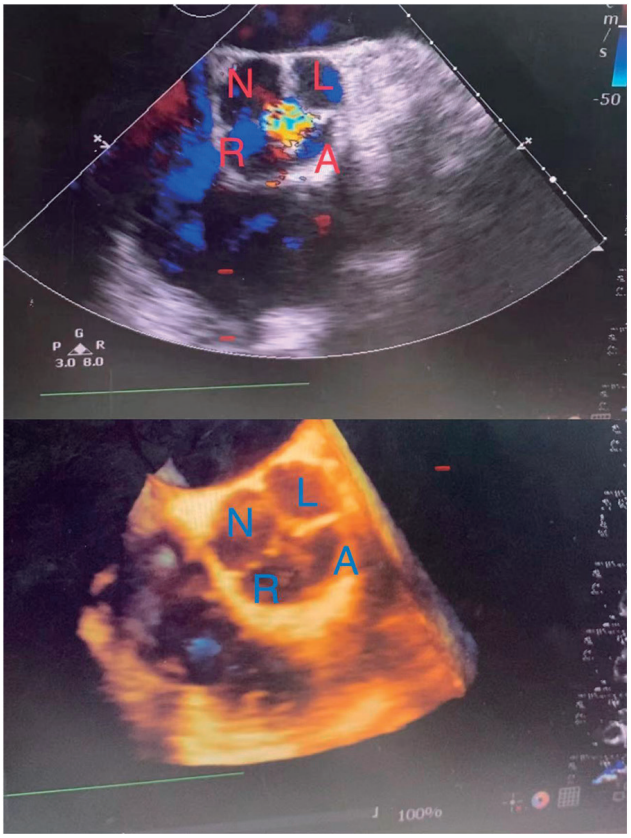

Fig. 1 Preoperative TEE found QAV with an accessory cusp between the left and the right coronary cusps. TEE: transesophageal echocardiogram; QAV: quadricuspid aortic valve

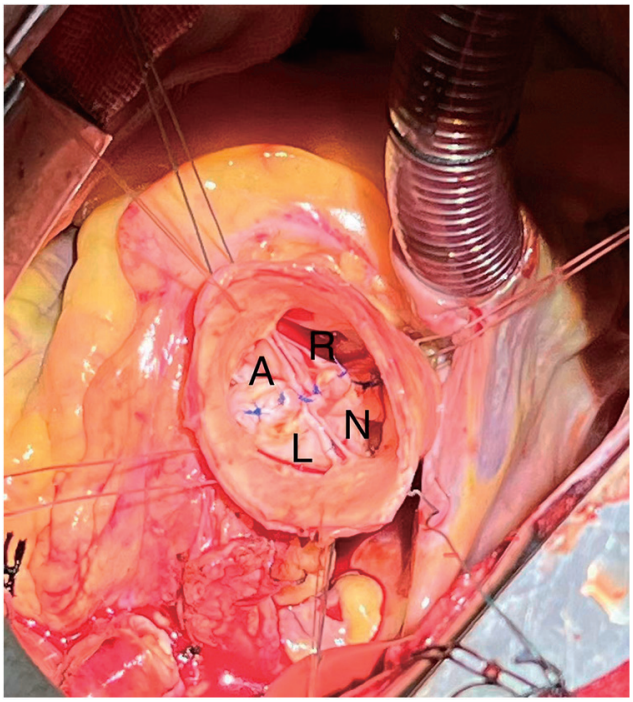

Fig. 2 Bicuspidized aortic valve by closure of the commissures between the left noncoronary and the right accessory cusps.

the later annuloplasty. Modification of the commissures was performed at first: the fusions between the left accessory and the right non commissures were carefully split off using a number 15 blade, the calcifications were removed with very delicate acts, the false chordaes were cut off thereafter, and the raphe formed by the false chordae was carefully shaved. On seeing the tissue was sufficient, the calcified part on the accessory cusp was removed by a triangular resection. Four subvalvular fixing points were then implanted symmetrically to stabilize the basal aortic ring without touching the cusps. The commissures between the larger and the smaller cusps were plicated using interrupt polyethylene (5-0) in order to minimize the chance of cusp retraction. The caliper (MSS-1; Fehling Instruments, Karlstein, Germany) was used to assure that the effective heights of the two neo-leaflets were over 8 mm. A number 18 Hegar could freely pass through the aortic orifice. A 2-mm wide Dacron ring (Terumo, Vascutek Limited, Inchinnan, Scottland, UK) of 30 mm was trimmed and cut up, passed through the subcoronary tunnel, and rejoined and fixed to the aortic basal ring by the four points prepared earlier. The ascending aorta was cut completely from 5 mm above the sinotubular junction level to 5 mm below the brachiocephalic trunk level and replaced by a conduit of 30 mm Dacron (**Fig. 2**) Postoperative TEE showed that the aortic annulus was 22 mm, the aortic valve was bicuspid, the effective height was 10 mm, the peak velocity of the valve was 2.17m/s, and the mean transvalvular gradient was 11.4 mmHg (**Video**; the video is available online.)

The patient was extubated on the operative day and discharged 6 days after the surgery without complications.

Discussion

Quadricuspid aortic valve (QAV) is not common. Idrees et al.[1] reported 31 QAV cases among 19722 patients who underwent aortic valve surgery in Cleveland Clinic from 1989 to 2010; only 7 of them had their native aortic valve repaired, suggesting that although this kind of abnormality is rare, successful repair is a great challenge.

To perform a successful aortic valve repair in QAV, preoperative considerations are very important. Correct measurements of aortic annulus, sinus of Valsalva, sinotubular junction, and ascending aorta can help us to determine which parts of the aortic root need to be repaired during the surgery. The Cleveland report[1]

revealed that more than half of the QAV cases presented dilatation of the proximal aortic diameter (>4 cm). In addition, QAV is commonly associated with anomalous origin of coronary arteries.[2] It is important for surgeons to be aware of any abnormality prior to the surgery.

The surgical strategy is another key for a QAV repair. Totally, the QAV can be morphologically classified into 3 types: type 1: 4 equal cusps, type 2: three larger cusps and one smaller accessory cusp, and type 3: two larger cusps and two smaller cusps. For patients of type 1 and type 3, bicuspidization is a reasonable option because the angle between the commissures is often 180 degrees, which can distribute the pressure equally and make the structure more stable as we do in aortic repair for patients of BAV. For those patients of type 2, tricuspidization may be the preferred technique: to eliminate the small accessory cusp by suturing it to the neighbor cusp or excising it completely to restore the tricuspid fashion of the valve.

We would like to discuss some pearls and pitfalls that we gained in our case:

1. Be sure of sufficient tissue before triangular resection on a cusp. It is very important to make precise measurements such as the geometric height before picking up the blade.
2. Be aware of the potential stenosis if the strategy of bicuspidization is decided, particularly when the sinus of Valsalva is not dilated. Excess plication of the cusps can lead to excess supravalvular acceleration postop.
3. Annuloplasty is always necessary for aortic valve repair. The role of annuloplasty in aortic valve repair is more and more recognized.[3] The later dilatation of the aortic annulus is an independent risk factor for recurrence of aortic regurgitation.
4. Sequence of the procedures is sometimes crucial for the success. Very frequently, aortic valve repair cannot be alone. Additional procedures such as annuloplasty, aortic root reconstruction, and ascending aortic replacement are needed for long-term durability. We should well organize the sequence of the procedures to avoid any potential injury to the cusps.

Conclusion

QAV can be repaired under deliberate preoperative considerations. Precise measurements, appropriate surgical strategy, and thoughtful organization can help to make the repair correct.

Informed Consent

We declare we have obtained the informed consent from the patient.

Disclosure Statement

All of the authors have no conflict of interest.

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