A huge saccular aortic aneurysm compressing right coronary artery 7 years after aortic valve replacement due to bicuspid aortic valve insufficiency

A rare case report

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Abstract

Background: In a patient underwent aortic valve replacement (AVR) due to bicuspid aortic valve (BAV) insufficiency without marked dilation of ascending aorta, the development of delayed-typed aneurysmal complication of ascending aorta has been often reported because the dilated aorta tends to grow insidiously with age.

Case summary: A 58-year-old man who underwent AVR with mechanical valve due to severe aortic regurgitation secondary to BAV 7 years previously presented with exertional chest discomfort for 1 year. An echocardiography showed a well-functioning aortic valve, and an MPR (magnification reformating, NYHA = New York Heart Association, RCA = right coronary artery.

Conclusion: After successful AVR in the patient with BAV insufficiency and mildly dilated ascending aorta, a regular aortic imaging such as cardiac MDCT with aortography would be helpful to monitor the morphology and size of ascending aorta and related complications to guide future management.

Keywords: Aortic valve replacement, BAV, multiplanar reformatting, NYHA = New York Heart Association, RCA = right coronary artery.

1. Introduction

Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly, occurring in 0.46% to 1.37% of the general population.[1] Aortic valve stenosis and regurgitation are the most frequent complications of BAV. Abnormalities of the aorta such as dilatation, bicuspid aortopathy, coarctation, cystic medial necrosis, and dissection are also often present in affected persons and can be potentially life threatening.[2-4] The prevalence of dilatation of the ascending aorta among persons with a bicuspid aortic valve is reported in anywhere from 20% to 84% of patients.[5] After successful aortic valve replacement (AVR) due to BAV insufficiency, the dilated aorta tends to grow insidiously with age compared to a normal tricuspid aortic valve.[6] Development of large, delayed-typed aneurysmal change following AVR has been previously reported.[6]

Hereby, we report a rare complicated case with an unusual type of aortic dilatation 7 years post-AVR that was performed in the setting of BAV insufficiency.

2. Case report

A 58-year-old man came to our outpatient department complaining of exertional dyspnea (NYHA class I or II/IV) and intermittent chest discomfort. The symptoms had developed insidiously over the past year. His chest pain and dyspnea had been getting progressively worse during the last 6 months. On physical examination, irregular heartbeats without significant murmurs were noted.

Seven years ago, the patient had initially presented to our clinic complaining of exertional dyspnea (NYHA class II/IV). An initial transthoracic echocardiography demonstrated a BAV with severe aortic regurgitation but without significant calcification. The echocardiogram also demonstrated normal left ventricular systolic function (ejection fraction: 60%) with eccentric hypertrophy and loss of A wave due to atrial fibrillation. The diameter of the aortic root was measured to be 4.56 cm at that time.
Invasive coronary angiography with aortography before surgery revealed no significant coronary luminal narrowing and a mildly dilated ascending aorta (Fig. 1A and B). He underwent successful AVR using a mechanical prosthetic valve (Saint Jude #25) without additional aortic surgery.

His current medications include warfarin 4mg for stroke prevention from atrial fibrillation. His antihypertensive regimen included fimasartan 30mg daily, nebivolol 2.5mg daily, and indapamide 1.5mg daily. He has been compliant with his medication regimen.

The patient’s most recent ECG demonstrated atrial fibrillation with controlled ventricular response. A chest x-ray showed no cardiomegaly. A follow-up echocardiography demonstrated a well-functioning prosthetic aortic valve and normal left ventricular systolic and diastolic function.

In order to evaluate the cause of dyspnea with chest pain, a cardiac multidetector computed tomography (MDCT) was performed. Two-dimensional coronary cross-section images revealed a large saccular-typed ascending aortic aneurysm (79.7 mm x 72.8 mm in size) just distal to the prosthetic aortic valve annulus (Fig. 2A and B). Curved multiplanar reformatted images also demonstrate diffuse, eccentric 90% luminal narrowing of the proximal right coronary artery (RCA) resulting from external compression by the aneurysmal sac. Also noted within the aneurysm was a low-density signal suggestive of mural hematoma or thrombi (Fig. 2C). Three-dimensional volume rendering images with coronary endoluminal and thrombotic settings confirmed compression of the proximal right coronary artery lumen by the aortic root aneurysm (Fig. 3A–F). The patient subsequently underwent open-heart surgery with resection of the ascending aortic aneurysmal sac and consecutive ascending aorta and hemi-arch replacement using a graft (Hemashield #28). The pathology of the resected aneurysmal sac demonstrated a vascular wall with myxoid degeneration and dystrophic calcification with organized thrombi. The patient’s post-operative course was uncomplicated, and he has been doing well in outpatient follow-up. Informed consent was given by the patient.

3. Discussion

In a patient with BAV insufficiency including severe stenosis or regurgitation, AVR is usually recommended instead of valve repair according to the current valvular heart disease guidelines. Three different types of ascending aorta dilatation in the patient with BAV are known as follows; Type 1 is most common type and characterized by dilatation of tubular ascending aorta (primarily along convexity of aorta) with mild-to-moderate root enlargement. The patients with type 1 have an older age at diagnosis, higher incidence of aortic stenosis, and the right-left cusps fusion pattern. Type 2 is associated with arch dilatation and involvement of tubular ascending aorta, with relative sparing of root. It is usually accompanied by the right-noncoronary cusps fusion pattern. Type 3 involves aortic enlargement confined to aortic root with normal tubular ascending aorta and arch sizes. This case demonstrates a saccular aneurysm which does not fall within the classic classification system based on the current literature. Furthermore, a large aneurysmal sac compressing the RCA appears to be a very rare and complicated presentation.

In patients with BAV and dilation of the aortic root or ascending aorta, surgical intervention to replace the aortic root and/or ascending aorta is determined by the aortic size according
to avoid life-threatening aortic dissection. If the diameter of the ascending aorta is >5.0 cm and the patient has risk factors for aortic dissection including rapid aortic growth rate (≥0.5 cm/yr) and familial history of aortic dissection or if a skillful surgical team performs the procedures to the patient at low operative risk, surgical correction of the ascending aorta is also reasonable even in asymptomatic patients. Furthermore, for patients with BAV undergoing AVR because of severe aortic insufficiency, replacement of the ascending aorta is recommended if the diameter of the ascending aorta is >4.5 cm based on expert opinion of current guidelines. In these cases, close evaluation of the morphology and size of ascending aorta should be performed before operation.17–10–13

After successful AVR without replacement of the ascending aorta due to any cause in patients with BAV, aortic magnetic resonance angiography or computed tomography such as chest or cardiac MDCT with aortography is recommended to monitor the size and morphology of the ascending aorta. These evaluation should be performed annually especially in patients with an aortic diameter >4.5 cm after successful AVR according to expert recommendations.12

This case suggests that the use of aortic and coronary imaging using cardiac MDCT may be helpful in elucidating less common causes of vague or unexplained chest discomfort and dyspnea in the absence of significant abnormalities on echocardiography after aortic valve surgery. This imaging modality may also help predict or prevent lethal aortic complications.

4. Conclusion

We present a rare case of huge saccular aneurysm compressing the RCA 7 years after AVR in a patient with BAV insufficiency. In a case of AVR without surgical repair or replacement of the ascending aorta due to any cause, cardiac MDCT with aorta and coronary angiography would be helpful to monitor the morphology and size of ascending aorta and to guide future management.

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