Delayed aneurysmal complication of bicuspid aortic valve disease after heart transplantation

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ADULT: TRANSPLANTATION: CASE REPORT

CLINICAL SUMMARY

A 72-year-old man with bicuspid aortic valve (BAV) disease underwent aortic valve replacement (AVR) in 1993, and subsequently developed cardiomyopathy secondary to sarcoidosis for which he underwent orthotopic heart transplantation in 1995. Posttransplantation, he developed mild hypertension in the range of 130 to 140 mm Hg (systolic), but otherwise was doing well clinically and undergoing routine echocardiographic surveillance. In late December 2017, he presented to an outside hospital with chest discomfort. Echocardiography at that time showed normal function of the transplanted heart and an ascending aortic diameter of 3.6 cm. As part of a workup for presumed pneumonia, he underwent a computed tomography (CT) scan of the chest, which incidentally showed a distal ascending aortic pseudoaneurysm of the native ascending aorta directed posteriorly between the right pulmonary artery and superior vena cava. The donor aorta was nonaneurysmal and not involved in the disease process. After careful dissection, the ascending aorta was freed and a crossclamp was applied. The aneurysm was resected and transverse hemiarch replacement was performed under deep hypothermic circulatory arrest with retrograde cerebral perfusion. Upon completion of the distal anastomosis, cardiopulmonary bypass was resumed, and ascending aortic replacement was performed (Video 1). Follow-up clinic visit at 1 and 6 months showed stable repair by CT imaging (Figure 1).


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DISCUSSION

To our knowledge, this is the first reported case in the literature of a recipient native ascending aortic aneurysm in a patient with BAVV disease after cardiac transplantation. Other reports have described thoracic aortic aneurysms in the recipients’ native aorta unrelated to BAVV disease, or complications of the donor aorta associated with BAVV disease in the donor heart. Nevertheless, a retrospective review of 243 explanted hearts found that 7 patients had native (recipient) BAVV disease, suggesting that the prevalence of BAVV disease in the heart transplantation population may be as high as 2.8%. The etiology of this aneurysm is likely a combination of risk factors related to both BAVV disease and heart transplantation. Current theory suggests that ascending aortic dilatation in BAVV disease results from hemodynamic and embryologic influences. Because any hemodynamic abnormalities associated with the morphologically bicuspid valve should have been eliminated at the time of this patient’s AVR, this case underscores the importance of the embryologic component of aortopathy in BAVV disease. Aneurysm formation is also more prevalent after heart transplantation than in the general population due to factors unrelated to BAVV disease. For example, immunosuppression may directly increase the rate of atherosclerosis and weaken aortic wall tissue. The patient was taking prednisone (5 mg daily), which has been associated with aneurysm expansion in animal models, and tacrolimus (0.5 mg twice per day), which is known to induce hypertension. Moreover, hemodynamic changes related to improved cardiac output after heart transplantation could exacerbate the problem.

Although the exact cause for aneurysm formation in this patient may not be known, this case demonstrates the necessity for follow-up thoracic aortic imaging in BAVV patients. The 2018 American Association for Thoracic Surgery guidelines for BAVV aortopathy recommend regular interval imaging of the thoracic aorta regardless of baseline diameter to screen for aneurysm formation. Even in patients with normal aortic dimensions, the thoracic aorta should be imaged at least every 3 to 5 years (if stable). Furthermore, these recommendations apply to BAVV patients even after AVR because of the continued risk of aortic complications. At our institution, surveillance CT imaging is not a routine part of follow-up care after heart transplantation. Therefore, future studies should assess the need for surveillance imaging of the thoracic aorta in patients after heart transplantation, especially in the presence of BAVV disease and other risk factors for aneurysm formation.

CONCLUSIONS

Surgery on the ascending aorta can be done safely with minimal surgical complications many years after cardiac transplantation. Routine surveillance with follow-up CT imaging of the thoracic aorta may be warranted in BAVV patients after cardiac transplantation.

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