Aortic valve replacement in a diseased bicuspid valve eleven years after transplantation

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Abstract

Cardiac allotransplantation is subject to a number of chronic complications that may limit graft survival. These include allograft coronary artery disease, renal dysfunction, hypertension, and malignancy, which are largely due to the immuno-modulatory and adverse effects of transplant medications. Reoperation for native allograft disease progression is a rarer phenomenon. We report a case of aortic valve replacement for bicuspid aortic valve stenosis that occurred in a patient more than ten years after cardiac transplantation.

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1. Clinical summary

A 55-year-old gentleman underwent orthotopic cardiac transplantation in 1996 for idiopathic cardiomyopathy. The donor was a 42-year-old white female with an echocardiogram that showed good systolic function (LVEF > 50%) but an aortic valve that appeared to be bicuspid. In spite of this, there was neither stenosis nor regurgitation demonstrated on echocardiogram. Eleven years after transplantation, the patient presented with several episodes of photopsia associated with lightheadedness and near syncope. These episodes were not exertion-related and resolved after 20–30 s. Otherwise, he was entirely asymptomatic from a cardiopulmonary standpoint and without evidence of myocardial ischemia or congestive failure. Further workup included an echocardiogram that demonstrated an EF of 50–55% with an aortic valve mean gradient of 29 mmHg, peak gradient of 51 mmHg, and mild to moderate aortic regurgitation. Additionally, the bicuspid valve was severely calcified with exuberant calcification protruding into the left ventricular outflow tract. A preoperative catheterization revealed coronary arteries that were without focal disease. Based on these findings, the patient was taken to the operating room for aortic valve replacement with a 23-mm Edwards pericardial prosthesis. Operative findings confirmed a bicuspid valve that was extensively calcified and immobile. Pathology confirmed myxoid degeneration and calcifications. Postoperatively the patient followed an uneventful course and was discharged from the hospital on post-op day six.

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2. Discussion

Cardiac transplantation is performed worldwide more than 4000 times each year with a survival rate after one year that declines at a linear rate of about 3.4% even well beyond 15 years post-transplant [1]. Long-term survival is primarily limited by the development of allograft coronary artery disease (ACAD) which represents the most common cause of mortality after the first year. Given the lack of suitable donor organs available to support the increasing demand for transplantation among patients with end-stage heart failure, attention has turned to increased utilization of marginally acceptable hearts. Use of appropriate thoracic and cardiovascular interventions has been shown to improve long-term survival after heart transplantation. Traditionally, organic valve disease in the donor heart has been considered an absolute contraindication to cardiac transplantation. However, there are case reports of successful outcomes in patients who underwent mitral valve repair either at the time of transplantation or in the post-transplant period [2, 3]. To date, there have only been a small number of cases in which aortic valve replacement was required in the donor organ. In one case, concurrent aortic valve replacement and coronary artery bypass grafting were performed at the time of transplantation [4]. In another case, endocarditis mandated replacement of the allograft valve after transplantation [5]. Aortic valve replacement of a regurgitant donor valve has been reported in a patient with a bicuspid valve 31 months after transplantation [6].

Given the limited donor supply and the improving outcomes in patients with marginal organs, meticulous screening has become of paramount importance. Using an alternate list for marginal donors has been proposed as a
strategy for increasing the yield of available organs. Although pre-existing valvular disease was not included in the list of donor criteria for assignment to the alternate list, Felker and colleagues demonstrated improved outcomes relative to medical therapy in end-stage heart failure patients [7]. In a consensus statement on maximizing the use of donor organs, Zaroff et al. note that while echocardiography is effective in the assessment of anatomic abnormalities, this strategy fails to reliably predict functional outcomes [8]. In many cases, right heart catheterization is necessary to determine the appropriateness of an organ for transplantation. Bench-top interventions also have the potential to improve marginal organs into suitable donors as in cases of tricuspid regurgitation, other mild valvular abnormalities, or secundum-type atrial septal defects.

This case also raises the question of how often to screen a transplant patient with known valvular abnormalities for the development of surgical pathology. The natural history of asymptomatic patients with a bicuspid aortic valve in the absence of hemodynamic compromise demonstrates excellent long-term survival. However, Michelena and colleagues recommended the use of routine echocardiographic assessment in patients with valve degeneration [9]. Given the hemodynamic and inflammatory disturbances often associated with cardiac transplantation, it seems reasonable to closely follow patients with known anatomic variants in the same manner. This patient was followed with echocardiograms every three months for the first three years, followed by every six months for two years, followed by annual studies. Although there is not enough clinical experience following recipients with anatomic variants to make an evidence-based recommendation, this conservative surveillance schedule seems reasonable, given that it enabled detection of this diseased valve in a timely manner. While most centers practice routine echocardiography in the follow-up of their patients after cardiac transplantation, there are currently no established practice guidelines for this screening modality. We feel that this case lends support to the aggressive strategy employed at the Johns Hopkins Hospital as outlined above.

In contrast to these previously published cases of donor heart valve disease, this patient’s disease process was not clinically evident until eleven years following transplantation. He required aortic valve replacement for progressive organic valvular disease. This case demonstrates both the survival benefits of transplantation with a marginal donor organ over optimal medical therapy as well as the increasing ease with which interventions can be performed for patients with multiple prior surgeries. It lends support to the concept of valve surgery following transplantation and underlines the importance of meticulous donor organ screening prior to transplantation.

References