CASE REPORT

Late aortic dilatation and regurgitation after Ross operation

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Abstract

The Ross operation, a procedure of replacement of the diseased aortic valve with an autologous pulmonary valve, has many advantages such as no need for anticoagulation therapy and similar valve function and growth potential as native valves. However secondary aortic disease has emerged as a significant complication and indication for reoperation. We report a 48-year-old woman who had Ross operation in 1997 for a damaged bicuspid aortic valve and severe aortic regurgitation due to subacute bacterial endocarditis complicated by aortic root abscess. In 2009, 12 years later, progressive severe aortic regurgitation with incomplete coaptation and mild dilatation of the aortic root was shown on echocardiography and contrasted CT, while the pulmonary homograft retained normal function. She subsequently underwent aortic valve replacement. Histopathological examination of the explanted neo-aortic valve and neo-arterial wall revealed pannus formation at the nodulus Arantii area of the three valve cusps, ventricularis, and arterialis. The amount of elastic fibres in the neo-aorta media was less than usual for an aorta of this patient’s age but was similar to a pulmonary artery. The pathological findings were not different from other studies of specimens removed between 7 to 12 years after Ross operation. However, the pathophysiology and long-term implications of these findings remain debatable. Considering the anatomical and physiological changes induced by the procedure, separate mechanisms for aortic dilatation and regurgitation are worthy of consideration.

Keywords: heart valve prosthesis implantation, aortic valve, aortic valve insufficiency, cardiac pathology

INTRODUCTION

Ross operation involves replacement of a diseased aortic valve using the autologous pulmonary valve and implantation of a homograft at the removed pulmonary valve site.¹ The most important characteristic of the Ross operation is the use of the pulmonary autograft instead of a homograft which has many advantages, among them, the availability of appropriately-sized grafts, almost permanent life-span of the valves and less probability of infection. Also, several studies have shown that the patients do not require anticoagulation therapy and that the function and growth potential of autografts are similar to native valves.²⁻³ Ross operation is now broadly applied to patients with congenital and acquired aortic valve disease such as aortic stenosis, bicuspid aortic valve, endocarditis involving the aortic valve, and complex left ventricular outflow obstructive disease.¹² Long-term follow up data of Ross shows that the original subcoronary technique is rarely associated with autograft dilatation.¹ However, the freestanding technique developed later has resulted in a significant increase in aortic root disease as the most common complication and also the most significant indication for reoperation these days.⁴ There are a few reports on the pathological features of neo-aorta and neo-aortic valves⁵,⁶ but the long-term pathological effects on the valve and the wall after Ross operation has still to be elucidated. We report the clinicopathological features
of a patient with aortic root dilatation and regurgitation 12 years after Ross operation.

CASE REPORT

The patient was a 48-year-old woman, who had Ross operation in 1997 to correct subacute bacterial endocarditis on a bicuspid aortic valve complicated by aortic root abscess and severe aortic regurgitation. The pulmonary valve and trunk were dissected and implanted at the aortic site and the coronary arteries are reimplanted. Her pulmonary valve was replaced with a 24mm homograft. After the operation, patient was followed-up every 4-6 months. Follow-up transthoracic echocardiography showed progressive severe aortic regurgitation, with grade increasing from I-II in November 2000 to III-IV in January 2009. Also, the left ventricle dimension was slightly increased from 53/36mm in November 2000 to 58/44mm in January 2009. There was no episode of infection or malfunctioning of the autograft valve. The pulmonary homograft showed normal function until now.

Signs of progressive regurgitation of the aortic valve were noted from complaints of progressive dyspnea (NYHA Fv II-III/IV) from January 2009. Supporting information was obtained from preoperative echocardiography performed on March 2009, such as diameters at several points of the aortic root: aortic annulus 21.6mm, sinus of valsalva 37.6mm, sino-tubular junction 39.6mm, ascending aorta 42.9mm, aortic valve 4.8cm. There was severe aortic regurgitation with incomplete coaptation and mild dilatation of the aortic root, while the pulmonary homograft functioned normally (Fig. 1). The left ventricle was mildly dilated and showed diffuse hypokinesia of wall motion. The estimated ejection fraction was 48~53%. The mitral valve was thickened with flow acceleration. Both mitral and tricuspid valves showed mild regurgitation. On CT with contrast and 3-dimensional reconstruction of the image, there was aortic root dilatation with left ventricle dilatation resulting in cardiomegaly (Fig. 2).

Reoperation for aortic valve replacement with a St. Jude 21mm graft was performed in April 8, 2009. The removed neoaortic valve was collected (Fig. 3) and subjected to histopathological examination.

There was no specific event in the post-operation course. Simple chest X-ray showed no obvious aortic dilatation remained. The patient is on warfarin medication and being followed-up on an outpatient basis.

Pathology

Histopathological studies were performed on the removed neo-aortic valve which was originally a pulmonary valve and the removed neo-aortic wall which was originally the pulmonary trunk. Masson’s trichrome and elastic stains were used to reveal histomorphological changes.

There was a significant amount of pannus at the valve (Figs. 3 & 4). Most significantly, nodular fibrous growth was noted at the nodulus Arantii area of the three valve cusps. This nodular pannus was continuous with an additional pannus at the ventricular side of the valve, which was a thin layer of fibrous tissue covering the ventricularis. Pannus at the arterial

Fig. 1: Echocardiograms (parasternal long axis view) before reoperation. A. Left ventricle (LV) and aortic root (AO) shows dilatation. Also mitral valve leaflets are thickened. B. On Doppler sonography, blood flow (blue colored) is regurgitating through the aortic valve. There are mild regurgitation of mitral valve, too. LA, left atrium; RV, right ventricular infundibulum.
side, which is histologically called arterialis or fibrosa was also noted in some areas to a less significant degree.

The pathology of the neo-aorta was only a mild degenerative change (Fig. 5). Some foci of loss of elastic fibers were noted but to an insignificant degree. The amount of elastic fibres in the media of the neo-aorta was less than the usual aorta for the patient’s age but was similar to or slightly more than a pulmonary artery. Fibrosis of the media was not found and there were no signs of occlusion of the vasa vasorum or fibrous adhesion of the adventitia.

DISCUSSION

Histological findings of explanted pulmonary grafts

The first systematic study about pathological findings of pulmonary autografts was published in 2004 by Rabkin-Aikawa et al. They compared normal pulmonary and aortic valves with early and late pulmonary autografts at 6 years after Ross operation. Valves of the pulmonary autografts showed near-normal 3-layers with intact elastin and collagen structures. There was intimal thickening (pannus formation) on the ventricular aspect of some cusps. They also observed that aortic walls had a variety of findings including loss of medial smooth muscle cells and elastin, and variable granulation tissue in early explants. Variable disruption of structure, scarring, or both without inflammation or calcification were observed in late explants. The authors however, did not detail out which late autograft cases had aortic dilatation/regurgitation or not. So it is unclear whether the histological findings were due to normal adaptation of
Pathological abnormalities of dilated pulmonary autografts were studied by Takkenberg et al. in 1999. The arterial wall in a patient with aortic regurgitation and dilatation of the aorta showed focal interruption of the media with total absence of elastin fibers and intimal proliferation with fibrosis. In 2007, Dixit et al. reported a case in which the patient underwent reoperation for aortic regurgitation with mild dilatation after Ross operation 9 years previously. The explanted pulmonary valve cusps showed thickened layers of pannus on both the flow and non-flow surfaces. Immunohistochemical staining for SMA (Smooth Muscle Actin) confirmed the presence of proliferating smooth muscle cells. Large deposits of acid mucopolysaccharides in the zona fibrosa and of laminated elastic fibres in the pannus with papillary fibroelastomas were also noted. The arterial wall showed complete loss of cellularity, identifiable elastic laminae, and focal fibrosis. There was marked fibroelastosis of the intima.

Our case was a late event, being 12 years after the Ross operation, and the basic pathological features of the aortic valves were similar to previous studies. This case suggests that pannus formation at the valves may be a result of hemodynamic events denying the architectural or intrinsic limitation of the pulmonary valve to be used for the aortic valve. It is important to note that these lesions are at least non-progressive.

It is not clear whether the minor degenerative changes of the media at the wall of the neo-aorta were the cause or result of dilatation of the aorta. But it is fair to say that there was no evidence of inflammatory aortic lesions as seen in various conditions like Behcet disease or ankylosing spondylitis. Vasa vasorum function is claimed to have a significant role in the integrity of aorta but our case did not reveal any adverse impact on the vasa vasorum by the Ross procedure.

The mechanism of late aortic root disease after Ross operation

Frigiola et al. reported echocardiographic follow-ups of patients after Ross operation and compared...
LATE AORTIC DILATATION AFTER ROSS OPERATION

The early reoperation cases with late reoperation ones. They noted that the 4 cases with early reoperation (<4 years) had rapid and severe aortic regurgitation without aortic dilatation, but the other 4 cases with late reoperation had definite aortic dilatation, leading to gradual aortic regurgitation. So they supposed that the former cases had technical problems at the time of operation which caused suboptimal results, while the latter may have a different mechanism related to the pathophysiological effects of the Ross operation itself.

There are three techniques in the Ross operation for adult patients, such as subcoronary freehand valve grafting (the original one), freestanding complete aortic root replacement (the current mainstream) and cylinder inclusion. Ross’s first long-term follow-up report based on 20 years experience, published in 1992, stated that no aortic regurgitation was found. Recently Sievers et al. confirmed again the advantage of the subcoronary technique Ross originally suggested. However, complexity of the subcoronary technique and frequent mismatch of aortic and pulmonary roots (particularly in infants and young children) prompted many clinicians to try more reproducible and successful variations.

The freestanding aortic root replacement, which requires mobilization and implantation of both coronary arteries as practised in the current reported case, became the most popular technique. However, it resulted in an increased prevalence of aortic root disease leading it to be the most common complication of Ross operation. The incidence rates vary depending upon the definition of aortic dilatation, such as size or location. The overall freedom from reoperation rate at 10 years ranges from 70% to 90% according to centres, mainly reduced by autograft insufficiency mostly related to late dilation of the autograft annulus.

So far, the freestanding root technique is suspected as the cause of aortic dilatation or regurgitation requiring subsequent reoperation, because these complications are rare with the subcoronary technique. One possible mechanism could be the over-dissection of the vasa vasorum which supplies the pulmonary arterial wall, leading to transmural ischemic.

Fig. 5: Microscopic images of arterial wall of pulmonary autograft. (A, C: silver stain; B, D: Masson’s trichrome stain) A --- B --- C --- D --- *, lumen; l, intima; M, media; A, adventitia
injury of the arterial wall. The freestanding technique requires more manipulation of the pulmonary arterial wall than the subcoronary technique, during autograft dissection, wall size adjustments, and implantation of the coronary arteries. This explanation has been suggested since 10 years ago. In an animal experiment, arterial wall removed vasa vasorum showed necrotic change of outer layers of the media with complete loss of smooth muscle cells, relatively preserved elastin fibres and their sheaths and close apposition of the elastic lamellae. These findings are consistent with former studies on dilated pulmonary grafts. Stefanadis et al. suggested that weakening of the media by these structural changes would result in disturbed absorption of hemodynamic forces that act on the wall, which in turn eventually lead to aortic dilatation.

Furthermore, the freestanding root technique requires implantation of pulmonary arterial roots. This procedure results in sudden exposure of the neo-aortic root to high pressure blood flow ejected from the left ventricle. With time, persistent exposure to high pressures result in histomorphological changes in the neo-aortic root.

Physiological mechanisms have very important roles in dilatation of the aorta and valve regurgitation. They are generally understood as being closely related. Cases with preoperative regurgitation of the aorta will have more chance of aortic regurgitation after Ross operation.

However, separate mechanisms may be responsible for aortic dilatation and valve regurgitation in some cases. The ascending aorta after Ross operation may be intrinsically defective due to its pulmonary origin. Dilatation of the neo-aorta may to some extent be an adaptive process rather than a pathological condition. It has been revealed that even normally functioning pulmonary autografts also dilate to a significant range. Dynamic differences have also been observed between pulmonary and aortic roots in animal experiments. Expansion of the pulmonary root commissural and sinotubular area showed similar dynamics, whereas the aortic root was significantly different. The dynamic features of the pulmonary root might explain global neo-aortic root dilatation, particularly at the commissural and sinotubular junction levels. The pulmonary root is much more distensible within its normal pressure range (0-30 mmHg), but as the pressure increases further, it can hardly dilate beyond the pressure of 100 mmHg because of limitation of distensibility. This 2-phase dilatation of the pulmonary root suggests that the root was working under abnormal conditions with loss of functional elasticity in the systemic range 80 to 120 mmHg.

The long-term implications of these findings are unknown, especially the combined effects of ischemic injury and long-term systemic pressure load. However, it has been suggested that the use of a shorter segment of pulmonary trunk over the sino-tubular junction would have less probability of aortic dilatation. An additional suggestion was usage of the original aorta at the convexity or right side of the ascending aorta and autograft pulmonary tissue at the anterior and left of the aorta so that the jet of aortic flow through the valve will impact on the original aortic tissue instead of pulmonary tissue.

Conclusion

We report a patient with dilatation of the aorta and regurgitation of the aortic valve at 12 years after Ross operation. Two mechanisms of aortic root changes after Ross operation have been suggested: ischemia of arterial wall induced by manipulation of vasa vasorum during freestanding technique and progressive exposure to systemic blood pressure. Our 12-year-specimen showed histological features consistent with previous reports of cases explanted earlier, such as 9 years and 7 years after Ross operation. It has been reported that neo-aortic root dilatation normally occurs after Ross operation to a certain extent, without aortic regurgitation. Considering the anatomical and physiological changes induced by the procedure, separate mechanisms for aortic dilatation and regurgitation are worthy of consideration.

REFERENCES


134
LATE AORTIC DILATATION AFTER ROSS OPERATION


