Unexpected outcome (positive or negative) including adverse drug reactions

Outcome of mitral valve repair in a patient with systemic lupus erythematosus

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Summary

We present a 17-year-old woman with a 5-year history of systemic lupus erythematosus (SLE). The patient referred to us with the rheumatic involvement of the mitral valve and was treated with mitral valve repair by ring annuloplasty and anterior leaflet augmentation with autologous pericardium. Follow-up, however, disclosed precocious calcification of the valve, necessitating mitral valve replacement.

BACKGROUND

It is not clear that a reconstructive mitral valve operation is the procedure of choice for systemic lupus erythematosus (SLE). Be that as it may, conservative surgery does not alter the natural history of the disease and the progression of valve thickening and calcification. To date, no clear consensus has emerged as to whether these valves should be replaced or repaired.

CASE PRESENTATION

The patient first presented at the age of 12 years with a history of polyarthralgia and facial rash with increased antinuclear factor; a diagnosis of SLE was, therefore, made. She also had a history of rheumatic fever at the age of 13 years. Five years had elapsed before the patient was referred to us for mitral valve surgery, at which time, SLE was in remission and she was taking prednisone and azathioprine, and her renal function test was normal.

Preoperative echocardiography disclosed massive mitral insufficiency, moderate stenosis and severe tricuspid valve insufficiency associated with a dilated left ventricle. The systolic pulmonary artery pressure was 70 mm Hg. At surgery, there was no pericarditis and pleurisy. A mitral valve analysis revealed a dome-shaped and moderately thickened anterior leaflet with a markedly retracted posterior leaflet. Verrucous vegetations were not visible on the leaflets. All the chordae were thickened and shortened. The annulus was mildly dilated, but there was no calcification. The anterior leaflet was augmented with a patch of autologous pericardium, which had been treated with 0.6% glutaraldehyde. A size 32 Carpentier-Edwards mitral ring was implanted. In addition, ring annuloplasty with an Edward ring size 30 was performed for the tricuspid valve. Postoperative transesophageal echocardiography and pre-discharge transthoracic echocardiography TTE showed that repair had achieved good results. The patient was given rheumatic fever prophylaxis and regular medication for the SLE was continued after the operation. Six months later, the patient remained haemodynamically stable with no evidence of mitral and tricuspid

incompetence on echocardiography, but mitral valve had become more thickened.

Nevertheless, 11 months after surgery, the patient returned with recurrent dyspnea. Echocardiography revealed a thickened anterior leaflet with a large fenestration and an immobile posterior leaflet. The mitral valve had severe regurgitation. Blood cultures were negative. The mean transmitral gradient was 6 mm Hg, and the systolic pulmonary artery pressure was 65 mm Hg. In addition, there was extensive calcification of the mitral annulus and leaflets. Tricuspid valve had trivial regurgitation. She did not suffer episodes of acute rheumatic fever after the operation clinically.

The patient was submitted to reoperation. The autologous pericardial patch had shrunk and was detached from the anterior leaflet in the posterior edge (figure 1). The posterior leaflet was rigid and calcified. A further conservative operation being out of the question, the valve was excised and replaced with a size 27 Edward valve.

Figure 1 Intraoperative photography showed explanted anterior leaflet; the autologous pericardial patch had shrunk and was detached from the anterior leaflet in the posterior edge.

Cultures of the explanted mitral valve showed no evidence of infection. A histological examination of the valve specimen showed a thickened fibrotic tissue with massive infiltration of calcium. The patient's postoperative course was uneventful. One year after surgery, she was in New York Heart Association class II, in sinus rhythm. The left ventricular ejection fraction was estimated at 65, and the mean pulmonary pressure was 20 mm Hg. She is currently on anticoagulation and in good general condition.

OUTCOME AND FOLLOW-UP

The patient appears to be in good physical condition at 5-year follow-up, with no other related problems.

Discussion SLE is a multisystem disease that can lead to a widespread microvasculitis in multiple organs including the heart. In patients requiring a mitral valve operation, mitral valve incompetence has been attributed mainly to fibrosing valvulitis predominantly involving the ventricular surface of the leaflets and the adjacent normal endocardium, ruptured chordae tendineae with fibrosing valvulitis, ruptured chordae tendineae without fibrosing valvulitis, and anterior papillary muscle necrosis resulting in chordal rupture with associated fibrosing valvulitis.¹ It is noteworthy that our patient also had a history of rheumatic fever with the involvement of the mitral valve.

Thickening, stiffness, calcification and fibrosis of the leaflet tissue and chordae are not specific and are not distinguishable from rheumatic lesions.¹ Unfortunately, our case demonstrated that the enlargement of the leaflet with glutaraldehyde-preserved autologous pericardium, as described by Chauvaud *et al*,² is not appropriate because of the incidence of pericardial involvement in lupus, leading to early failure. Also, a conservative operation does not alter the natural history of the disease and the progression of valve thickening and calcification. The cause of the appearance of the calcified vegetation is not clear. It could be related to healed bacterial endocarditis or renal failure.

The results of valve repair in patients with a history of SLE have been widely controversial. Hakim *et al*³ reported two cases of valvular repair, one of which was successful and the other one ended up in reoperation after 3 years. They also reviewed the literature and found 36 reports of valvular surgery in patients with SLE. A case report by Kalangos *et al*⁴ presented the first case of valvular repair in patients with SLE. They followed up their patient for 1 year and recommended valvular repair as the procedure of choice in patients with SLE. They concluded that anticoagulation might be associated with higher risks in young patients who require prolonged steroid use and who frequently experience renal failure and undergo haemodialysis.

As Chauvaud² described, a 17-year-old patient who was treated with mitral valve repair. Nonetheless, the

follow-up visits revealed excessive calcification of the valve, necessitating mitral valve replacement with a cryopreserved homograft.

Mitral valve replacement has been reported to be safe by a handful of other authors.^{5 6}

Given the aforementioned facts and the young age of our patient, we opted for valvular repair. The procedure, however, resulted in failure, necessitating a second operation after 1 year, in particular, because of the autologous pericardium. Our experience showed that patients with SLE might not achieve the best result with valvular ring annuloplasty and anterior leaflet augmentation with autologous pericardium. Moreover, as a proportion of the patients with SLE need anticoagulation due to antiphospholipid antibody, the administration of this group of drugs might not impose extra-risk on these patients. Finally, we believe that applying valvular replacement in the first place in our patient would have been associated with better outcome, although her pericardium looked healthy at the time. Even if we decide to repair the valve, it is advisable that autologous pericardium be not used because SLE involves all components of the heart, including the pericardium, conduction system, myocardium, heart valves, and coronary arteries.¹

Learning points

- There is still limited experience with valvular repair and replacement in patients with systemic lupus erythematosus.
- However, our results and those in the existing literature are in favour of mitral valve replacement in such patients.

Competing interest None.

Patient consent Obtained.

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