Case Report

Cardiac Autotransplantation for Aortic and Mitral Valve Replacement in a Patient with Nephrogenic Systemic Fibrosis

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Adequate exposure is a prerequisite for open valve surgery. The mitral valve can rarely be very challenging to expose. We describe a redo double valve replacement in a patient with nephrogenic systemic fibrosis in whom exposure of the mitral valve was achieved with cardiac autotransplantation.

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Adequate valve exposure is a prerequisite for open valve surgery. While exposure of the aortic valve is rarely problematic, the mitral valve may be more challenging to expose. It is usually accessed through various approaches. Regardless of the approach, adequate exposure to the mitral valve is crucial to a successful valve repair or replacement.

Case presentation

A 55-year-old man, with a history of haemodialysis-dependent chronic renal failure, presented aortic prosthetic valve endocarditis, with *S. mitis*, 4 years after valve replacement for aortic sclerosis. He was given intravenous antibiotics for six weeks, after which inflammatory parameters normalised on blood examination and blood cultures became negative. However, the echocardiogram showed a large vegetation under the aortic prosthesis, severe mitral stenosis, pulmonary hypertension (60 mmHg) and a slightly decreased left ventricular ejection fraction of 45%. The coronary arteries were normal on catheterisation. He was scheduled for excision of the aortic prosthesis and of the vegetation, and mitral valve replacement.

Cardiopulmonary bypass was established through the right axillary artery and the right femoral vein. A full sternotomy was performed and, after adhesiolysis, the aorta was clamped. Retrograde cardioplegia was delivered and the aorta opened transversally. The aortic prosthesis and vegetation were excised. This procedure was followed by an incision in the interatrial groove, but visualisation of the mitral valve was impossible. The myocardium was very rigid and fibrotic, precluding any better exposure of the valve without increasing the risk of rupturing the heart. We proceeded to a transatrial transseptal approach and finally to a Guiraudon approach to the mitral valve, but visualisation of valve structures was extremely difficult. There appeared to be no option but to explant the heart and perform the aortic and mitral valve replacement *ex vivo*. Cardiac explantation was performed by transecting the aorta and pulmonary artery and the two *venae cavae*. Cold blood cardioplegic solution was administered intermittently into the coronary sinus during the period when the heart was *ex vivo*. Two mechanical prostheses were easily implanted with everted pledgeted sutures (Figure 1). Cardiac reimplantation consisted of repair of

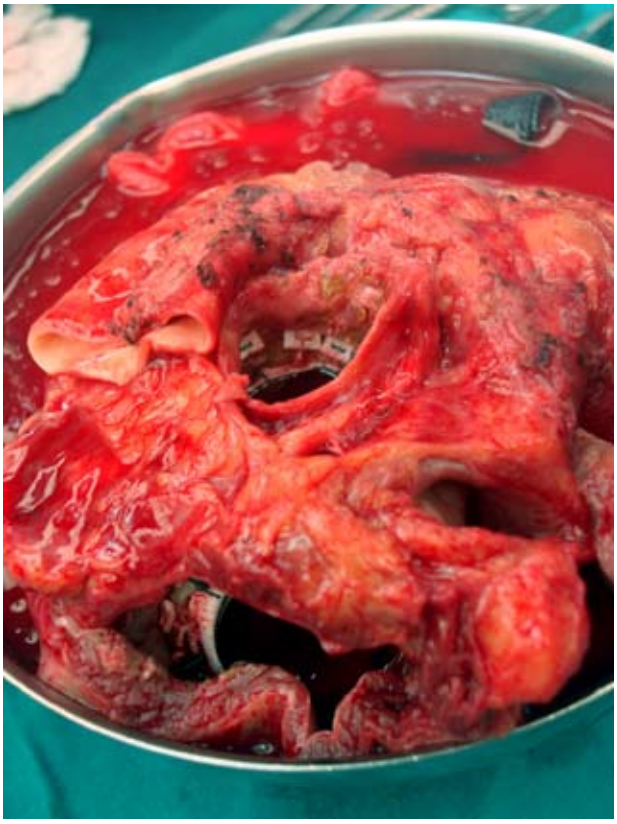

Figure 1. The aortic and mitral prostheses are already implanted.

the previously divided atria, and end-to-end anastomoses of the aorta, the pulmonary artery and the two *cavae* (Figure 2). Temporary epicardial pacing leads were placed. Total ischaemic time was 245 minutes, and the time on cardiopulmonary bypass was 335 minutes. The heart sustained good haemodynamics and, after full functional recovery, the patient was discharged home 7 days later. He is doing well four months after the operation.

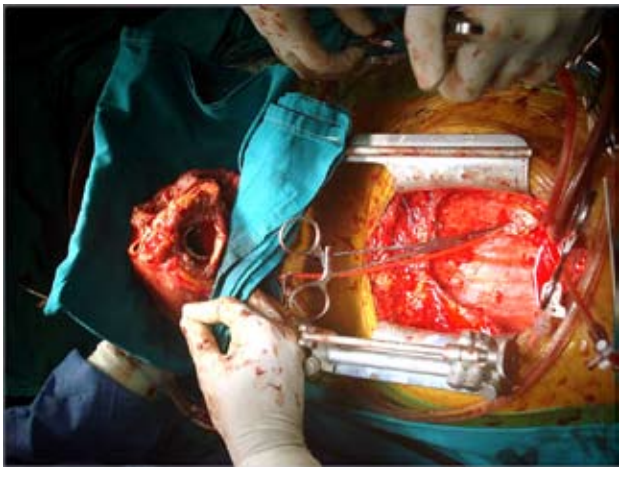

Figure 2. The heart is ready to be re-implanted in the patient's chest (head of the patient on the right of the picture).

Discussion

Exposure of the mitral valve is seldom difficult. However, after an experience similar to ours, Novitzky et al[1] did a poll on this subject, mailing a questionnaire to 3000 cardiac surgeons. Reponses were obtained from 1120. Inadequate mitral valve exposure had been experienced by 70%. To provide increased exposure, 50% had extended the initial atrial incision both horizontally and perpendicularly to the atrial groove, 17% had divided the superior *vena cava*, 1% had divided the inferior *vena cava*, and 1% had divided both *cavae*. Furthermore, 4% of surgeons reported being forced to abandon the operation in 71 patients because of inadequate exposure. Three hundred and twenty perioperative deaths were directly attributed to an incomplete surgical procedure. These results show that, despite the rarity of this problem, it carries a high mortality when it appears. Our patient was found to suffer from nephrogenic systemic fibrosis, previously known as nephrogenic fibrosing dermopathy. It is a novel fibrosing disorder characterised by prominent cutaneous and systemic fibrosis in patients with renal failure. This condition alters the elasticity of the myocardium, decreasing the visibility of the mitral valve.

Autotransplantation has previously been used as a method to treat permanent atrial fibrillation,[2] atrioventricular disruption,[3] or congestive heart failure.[4] It is obvious that this approach allows the surgeon to work on any aspect of the heart, irrespectively of the anatomy of the patient or the orientation of the lesion. Of course, this additional manoeuvre prolongs the time of the operation. For this reason, it is important to recognise quickly whether the operation can be achieved in the usual way or if *ex vivo* repair is necessary, in order to lose as little time as possible by trying different approaches. Unfortunately, it is very difficult to say how much the morbidity and mortality of the procedure increases with this approach. However, in our case we believe that the risk was not substantial, taking into account the devastating complications that could have arisen if we had persisted and not changed our strategy, or had been forced to abandon the operation.

In conclusion, we believe that explantation of the heart, with mitral valve replacement being performed *ex vivo* followed by reimplantation, should

be considered when access to the mitral valve proves impossible with standard exposure. To our knowledge this is the first reported autotransplantation for this indication. *Ex vivo* repair should also be considered promptly in patients under chronic haemodialysis who may suffer from nephrogenic systemic fibrosis or other infiltrating diseases, such as amyloidosis or haemochromatosis.

References

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