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Case report - Valves

Severe mitral regurgitation in acute eosinophilic endomyocarditis: repair or replacement?

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Abstract

Established eosinophilic endomyocardial disease usually causes progressive heart failure from diastolic restriction and/or valvular dysfunction. Surgical treatment typically involves resection of endoventricular fibrosis and atrioventricular valve replacement. However, little is known of the clinicopathological behaviour and perioperative course of this disease in the early stages. Mitral valve repair seemingly offers an attractive surgical option in this scenario as it avoids prosthetic thrombosis—a recognized late complication of this disease. This was attempted in conjunction with left ventricular thrombectomy in a patient with acute hypereosinophilia and congestive heart failure associated with severe mitral regurgitation and restrictive cardiomyopathy. Although the early clinical and echocardiographic outcome was excellent, the patient deteriorated rapidly after 3 months when identical pathology relapsed in the left heart causing recurrent severe mitral regurgitation and heart failure. Cardiac function improved markedly following a redo bioprosthetic mitral valve replacement. Prosthetic valve function remained satisfactory until death occurred 2 months later from refractory acute lymphocytic leukaemia. Rapid disease recurrence jeopardizing a conserved mitral valve in acute eosinophilic endomyocarditis cautions against surgical repair despite its many advantages. A bioprosthesis is associated with reduced thrombotic complications and may be the treatment of choice for this rare pathology. © 2004 Elsevier B.V. All rights reserved.

Keywords: Mitral valve repair; Thrombosis; Heart failure; Endocarditis

1. Introduction

The etiology of endomyocardial fibrosis (EMF), though much debated, is considered primarily immunological. A clinical form associated with systemic hypereosinophilia was described by Löffler in 1936 [1]. Progressive heart failure from restrictive cardiomyopathy and/or atrioventricular valvular dysfunction results in uniformly poor prognosis despite medical therapy. Surgical treatments for advanced EMF include heart transplantation and in selected cases endocardial decortication and valve replacement. Good long-term outcome has been reported with the non-transplant approach in chronic EMF [2]. In contrast, little is known of the clinicopathological behavior and perioperative course of acute eosinophilic endomyocarditis.

Furthermore, the role of valve-sparing surgery is not well defined in this context.

2. Case report

A 17-year-old man presented with a 1-month history of progressive exertional dyspnea (NYHA class II) and lethargy. Clinical examination revealed apical pansystolic murmur and a skin rash over the limbs. Hypereosinophilia ($> 54 \times 10^9/l$; upper limit: $0.4 \times 10^9/l$) was found prompting bone marrow biopsy demonstrating gross eosinophilic infiltration. Concurrent abnormalities included mild anemia (Hb 117 g/l) and polyclonal gammopathy. Mild asthma and previous parasitic gastroenteritis constituted relevant medical history. Echocardiography confirmed severe mitral regurgitation (MR), preserved left ventricular (LV) systolic function, restrictive cardiomyopathy with thickening of the LV inflow tract (LVIT) (20 mm), apical obliteration of

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the ventricles, pulmonary hypertension, abnormal endoventricular reflectance and probable LV mural thrombus. The posterior mitral valve leaflet (PMVL) with attendant chordae were adherent to the abnormal posterior LVIT (Fig. 1). Hypereosinophilic syndrome with cardiac involvement—Löffler's endomyocarditis (LEM)—was diagnosed. The patient was hospitalized, anticoagulated and commenced on cytoreduction therapy (corticosteroid, hydroxyurea and imitinab). Massive leucolysis precipitated acute myocarditis (troponin: 22 $\mu\text{g/l}$) with angiographically normal coronary arteries. Left ventriculography showed diffuse endocardial irregularities particularly around the LVIT. Heart failure progressed over the next 2 months with increasing hemoptysis despite aggressive medical treatment including α -interferon (15×10^6 U/day). In view of rapidly progressive LEM and stable hematology, the patient was referred for urgent surgery.

Through median sternotomy, aorto-bicaval cardiopulmonary bypass was instituted. The heart appeared externally normal. Following cardioplegic arrest, the mitral valve was exposed through left atriotomy. The anterior leaflet was normal but the PMVL and subtending chordae were tethered to the LVIT by layers of whitish-yellow material covering the entire posterior LV wall between the papillary muscles (Fig. 2). The entire PMVL was detached from the mitral annulus to facilitate thorough endocardial debridement. The relatively unaffected leaflet and chordae were then reattached to the annulus and reinforced with a 32 mm annuloplasty ring (Cosgrove-Edwards annuloplasty system, Edwards Lifesciences, Irvine, CA, USA). Satisfactory leaflet coaptation was demonstrated on LV distension. Through a right atriotomy, both tricuspid valve and right ventricle were found to be disease-free. On-table echocardiography confirmed satisfactory ventricular function with trace MR prior to routine closure.

Apart from persistent eosinophilia ($\sim 15 \times 10^9/l$), postoperative progress was uneventful. Supraventricular

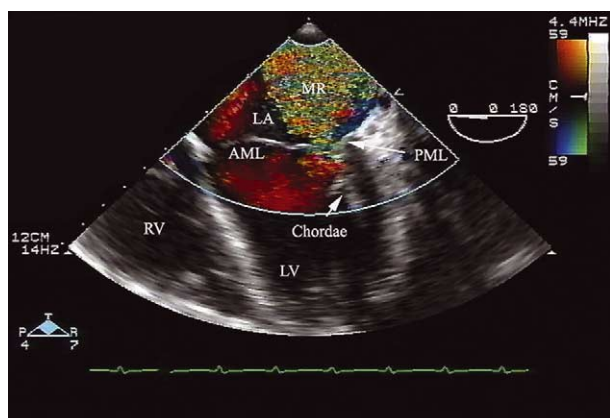


Fig. 1. Transesophageal echocardiogram demonstrating severe mitral regurgitation (MR) associated with a posteriorly based jet and tethering of posterior mitral valve leaflet (PML) and subtending chordae to the posterior annulus (LA, left atrium; AML, anterior mitral valve leaflet; LV, left ventricle; RV, right ventricle).

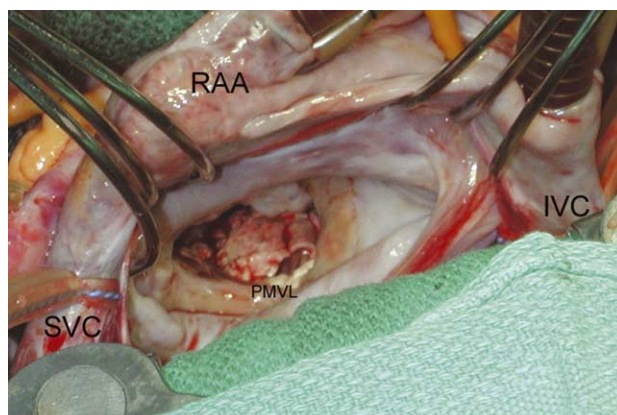


Fig. 2. Surgeon's view of the mitral valve through a standard left atriotomy demonstrating the abnormal thrombotic materials covering the posterior left ventricular inflow tract and the posterior mitral valve leaflet (PMVL) (RAA, right atrial appendage; SVC, superior vena cava; IVC, inferior vena cava).

tachyarrhythmia was successfully treated pharmacologically. The patient was discharged 12 days postoperatively in NYHA class I. Pre-discharge echocardiography documented satisfactory ventricular and valvular function. Histopathological examination of resected specimens confirmed laminated thrombus but without any trace of eosinophils embedded. Unfortunately heart failure recurred 3-months postdischarge leading to rehospitalization. Evaluation showed disease recurrence in the same sites producing severe MR. At reoperation, thrombotic materials again encased the PMLV and LVIT in addition to partial tears in the anterior leaflet. The annuloplasty ring was explanted and the valve excised and replaced with a 31 mm Mosaic™ stented bioprosthesis (Medtronic Inc, Minneapolis, USA). The postoperative course was uneventful except for persistent eosinophilia with the patient discharged after 7 days. However, he succumbed to acute leukemia despite intensive treatments 2 months later. Pre-mortem echocardiography confirmed unaltered cardiac and valvular function.

3. Discussion

Since Löffler first described 'endocarditis parietalis fibroelastica', other reports of a systemic illness characterized by refractory eosinophilia and multi-organ involvement followed [3]. Progressive cardiac failure results from restrictive cardiomyopathy and atrioventricular valvular dysfunction caused by EMF affecting one or both ventricles. The etiology is unknown although an immunologic basis was proposed based on links with inflammation, infection and allergy. Median survival, largely dependent on cardiac involvement, is ~ 24 months. Medical management has limited prognostic impact as heart failure is treated conventionally and cytoreduction therapy has little influence on endomyocarditis [3]. EMF without eosinophilia is also well recognized since first described in Africans

with heart failure. Some believe this idiopathic illness to represent a more chronic and milder variant of LEM. Apart from two discerning features, lack of systemic involvement and lower embolic potential in EMF, the two diseases share many clinical, radiological and hemodynamic similarities. Indeed the pathological findings in end-stage EMF and LEM are indistinguishable [4]. The persistent hypereosinophilia, skin rash, bone marrow and pulmonary infiltrates in our patient supported the diagnosis unequivocally. The clinical picture exemplified a rare disease with a male-predilection and rapid progression despite medical therapy. Although biventricular involvement is most common, the combination of severe MR and diastolic LV failure is well recognized in left-sided LEM.

Lepley first operated successfully on advanced EMF by performing endocardial decortication and mitral valve replacement (MVR) through an apical ventriculotomy [5]. This has subsequently been adapted effectively for right-sided disease. With 5-year survival of 72% achievable even amongst end-stage sufferers, surgery represents the only hope of prolonging survival in this incurable disease [2]. Keys to long-term success include thorough decortication and bioprosthetic valve replacement: xenografts constitute the bulk of reported experience whilst death from late thrombosis of a mechanical prosthesis has occurred [2,6,7]. Despite considerable surgical experience with managing the chronic fibrotic stages in LEM, the clinicopathological behaviour and perioperative course in the acute thrombotic stage remains a mystery. Our findings of extensive thrombosis involving the LVIT and PMVL correlated well with echocardiographic appearances and clearly explained the cardiac pathophysiology. This pattern resembled a previous case report in which fatal prosthetic thrombosis

occurred after MVR [7]. Mitral valve repair (MVR) has been described for advanced LEM but not in the acute stages [2, 8]. Although the valve-sparing approach potentially avoids late complications inherent with all prostheses, early postoperative recurrence of LEM suggests that MVR may be more appropriate.

In summary, redo MVR and LV thrombectomy was performed for congestive cardiac failure from early recurrence of LEM after MVR undertaken during the acute thrombotic stage. Valve-sparing approach should be avoided in this scenario particularly when eosinophilia persists.

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