

Mitral Valve Repair in Idiopathic Hypereosinophilic Syndrome

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Idiopathic hypereosinophilic syndrome is a rare systemic disease that can cause multiple organ failure by eosinophilic infiltration. Cardiac involvement is characterized by endocardial fibrosis and overlying thrombus, leading to restrictive cardiomyopathy and valvular dysfunction. Surgical experience of patients with mitral dysfunction caused by this syndrome is limited, and valvular replacement is most often performed. Mechanical valvular replacement has a high

incidence of recurrent obstructive thrombosis, and replacement with a bioprosthesis is recommended, despite associated restrictive cardiomyopathy. A patient is described who presented with mitral insufficiency associated with idiopathic hypereosinophilic syndrome, and underwent mitral valve repair.

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Idiopathic hypereosinophilic syndrome (IHS) is a rare systemic disease characterized by unexplained persistent eosinophilia associated with multiple organ involvement (1). The prognosis is correlated with heart involvement that results in a restrictive cardiomyopathy with endocardial fibrosis with/or mural thrombosis leading to valvular dysfunction (2). Surgical experience of patients with valvular dysfunction secondary to IHS is limited. Valve replacement is most often performed, but the choice between a mechanical or biological prosthesis in this setting poses a difficult problem. As mechanical valvular replacement has a high incidence of recurrent obstructive thrombosis, valve replacement with a bioprosthesis is recommended, despite association of restrictive cardiomyopathy.

The case is reported of a patient with mitral regurgitation caused by IHS who underwent mitral valve repair.

Case report

A 71-year-old man was admitted to the authors' hospital with congestive heart failure secondary to mitral insufficiency which had been diagnosed three weeks previously, with a first pulmonary edema. The

patient's previous medical history was notable for a moderate idiopathic eosinophilia diagnosed six months previously (eosinophil count $>1.5 \times 10^9/l$). On admission, the patient had a leukocyte count of $8.9 \times 10^9/l$ with 10% eosinophils ($0.89 \times 10^9/l$). This hypereosinophilia was considered to be idiopathic, as no other disorders known to cause secondary eosinophilia were found. No other organ dysfunction was associated with the condition.

Echocardiography revealed severe mitral insufficiency by restriction of motion of the posterior leaflet, without signs of endomyocardial fibrosis or thrombosis of the left ventricle. Right-heart catheterization showed a raised right heart pressure (pulmonary artery pressure 70/32 mmHg).

Surgical correction of the mitral insufficiency was performed through a sternotomy. The mitral valve was approached through the left atrial roof after wide dissection of the atrioventricular groove. Operative findings showed mural thrombus of the left ventricle involving the subvalvular apparatus of the posterior leaflet, leading to a restriction of leaflet motion. A thrombectomy of the left ventricle was performed through the mitral valve and completed via a transaortic approach. No underlying endomyocardial fibrosis was observed. The posterior leaflet was detached from the annulus from one commissure to the other in order to release the thrombus, which covered the ventricular face of the leaflet. A valvuloplasty associated leaflet extension with an autologous glutaraldehyde-treated pericardial patch and prosthetic annuloplasty with a

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32 mm Carpentier-Edwards Physioring® ring was then carried out.

Intraoperative transesophageal echocardiography revealed trivial residual mitral insufficiency with normal pulmonary artery pressure.

A histological examination showed the presence of eosinophils in the ventricular thrombus.

The patient's postoperative course was uneventful, and he was discharged having been prescribed warfarin for a three-month postoperative period. Since the patient's eosinophil count was normalized, specific treatment with hydroxyurea or prednisolone was not introduced.

At one year after surgery the patient remained asymptomatic, with no thromboembolic events. At the same time, echocardiography showed minimal mitral insufficiency without ventricular thrombus, and the eosinophil count was normal.

Discussion

Idiopathic hypereosinophilic syndrome is a rare disorder which is defined as an unexplained eosinophilia that lasts for more than six months and is associated with multiple organ dysfunction secondary to eosinophilic infiltration (1). Cardiac involvement occurs in more than 75% of patients with IHS, and is the major cause of morbidity and mortality in this syndrome. Cardiac manifestations include endocardial fibrosis with mural thrombosis formation, leading to restrictive cardiomyopathy and valve dysfunction by restriction of valve leaflet mobility (2). The valves most commonly involved are the atrioventricular valves with regurgitation and, less commonly, the aortic valve.

Mitral surgical experience in IHS is limited. In 1991, Boustany et al. (3) collected eight patients with IHS who underwent mitral valve surgery. Since that time, five other reports have been made, among which mitral valve replacement was performed most often

Table I: Surgical experience of mitral insufficiency in idiopathic hypereosinophilic syndrome.

Reference	Year	Procedure	Complication	Follow up	Outcome
Bell (4)	1976	Mitral annuloplasty	No	6 months	Alive
Weyman (5)	1977	MVR (bio) + tricuspid repair	No	6 months	Alive
Fauci (2)	1982	MVR+TVR (mech.)	Recurrent TE redo MVR, TVR (bio)	8 months	Died
		MVR+TVR (bio)	No	8 years	Alive
		MVR (bio)	No	4 years	Died
Blake (6)	1985	MVR (bio)	No	5 years	Alive
Hendren (7)	1988	MVR+AVR (mech.)	Thrombosis redo AVR, MVR (bio)	12 months	Alive
Boustany (3)	1991	MVR	Thrombosis	34 months	Alive
Inoue (8)	1991	MVR (bio)	-	-	Died post-op.
Arsiwala (9)	1995	MVR (mech.)	Thrombosis redo MVR	-	Died post-op.
Tamura (10)	1998	MVR (mech.)	-	-	Died post-op.
Radford (11)	2002	MVR (mech.)	Thrombosis redo MVR	8 years	Alive
Watanabe (12)	2002	MVR (mech.)	Recurrent thrombosis redo AVR, MVR (bio)	30 months	Alive

AVR: Aortic valve replacement; bio: Bioprosthesis; mech.: Mechanical prosthesis; MVR: Mitral valve replacement; post-op.: Postoperative; TE: Thromboemboli; TVR: Tricuspid valve replacement.

(12 patients), while only one patient underwent mitral valve annuloplasty (2-12) (Table I). The choice of valve prosthesis to be implanted in IHS patients is very difficult, as mechanical valves have a high incidence of obstructive thrombosis leading to reoperation with a high mortality (2,3,7,9,11,12). Activating eosinophils are able to modify coagulation and fibrinolysis by several effects which activate platelet aggregation (12). These effects are important during periods of uncontrolled eosinophilia.

Therefore, a bioprosthesis is recommended in the case of valve replacement, despite the frequent young age of patients and the associated restrictive cardiomyopathy with a small left ventricular cavity. Some authors have recommended additional anticoagulation, since small thrombi have been described on the bioprosthesis in IHS, despite treatment with warfarin. However, because of the rarity of the disease and the small number of patients who have undergone surgery, the long-term durability of bioprostheses in IHS is at present unknown.

For these reasons, mitral valve repair in IHS appears to be the treatment of choice. In this condition, the lesions are similar to those encountered in endomyocardial fibrosis that is reported in tropical areas. Mitral valve repair can be used in endomyocardial fibrosis in most cases if there is no calcification of the left ventricle and mitral valve. The repair in endomyocardial fibrosis consists of transvalvular endocardectomy after detachment of the posterior leaflet, often with the use of an autologous pericardial patch; this has led to good mid-term results (13).

The control of peripheral eosinophilia is imperative, with aggressive therapy consisting of prednisone, hydroxyurea or interferon. This medical therapy has led to an improved survival, with only 4% mortality after three years compared with 77% mortality within three years in the absence of such therapy. The complications of valvular surgery are also reduced (9).

Idiopathic hypereosinophilic syndrome is a rare cause of mitral insufficiency, yet mitral valve replacement in this setting is associated with a high rate of complications. Mitral valve repair can be performed, and appears to be the treatment of choice, but the procedure must be associated with medical control of peripheral eosinophilia.

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