Case Report

Heart Transplantation in a Patient with Endomyocardial Fibrosis

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Endomyocardial fibrosis is a common disease in tropical countries, particularly Uganda and Nigeria. It may affect the right and left ventricles, creating restrictive heart failure, which, in addition to the classical symptoms, manifests as ascites disproportional to peripheral edema. We report the case of a female patient with endomyocardial fibrosis refractory to clinical treatment, who underwent surgical treatment with clinical improvement for a short period. Three months after the surgery for resection of the endomyocardial fibrosis and tricuspid valvuloplasty, the disabling symptoms reappeared. Then the patient underwent bicaval orthotopic heart transplantation, which had a good clinical outcome. This was the first case of heart transplantation for the treatment of endomyocardial fibrosis, which proved to be a promising alternative.

Endomyocardial fibrosis is a common form of heart disease in tropical countries, such as Uganda [1] and Nigeria [2], and a lower incidence of it may be found in subtropical and tropical regions, such as Brazil, India, Colombia, and Sri Lanka. It is more common in the underprivileged population. It may affect the left ventricle alone (40% of cases), the right ventricle alone (10%), or both (50%) [3]. In the 2 latter situations, manifestations of right heart failure predominate. Clinically, the patients have huge ascites, little or no peripheral edema, even when left heart failure predominates. Patients with huge ascites are usually hard to clinically control and only slightly responsive to diuretics [4]. Some authors have suggested the existence of an inflammatory component in the formation of ascites [5]. Approximately 60% of the patients have eosinophilia [6] in the peripheral blood, which may be related to parasitic infestation.

The clinical treatment is symptomatic and offers little satisfaction [7]. When the disease reaches a more advanced stage, surgery provides symptomatic improvement, being then, the treatment of choice [3]. The best surgical results are obtained when the manifestations of left heart failure predominate [2,8,9]. The advanced disease has a poor prognosis, with a 2-year mortality rate ranging from 35 to 50% [10]. In those cases, heart transplantation may be a therapeutic option, although little used, with only one report of heterotopic transplantation reported in the literature.

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Case Report

The patient is a 50-year-old white female, housewife, born in the town of São Sebastião, in the state of Alagoas, who was referred to InCor in 1996, complaining of enlargement of the abdominal volume associated with lower limb edema for 3 years. The patient reported dyspnea associated with huge ascites, which improved with relief punctures performed every 20-30 days, even though she was using diuretics. The patient underwent investigation for hepatopathy in the gastroenterology unit of the Hospital das Clínicas. Once the viral, alcoholic and schistosomal etiologies were excluded, the patient was referred to InCor with the diagnostic hypothesis of chronic hepatopathy secondary to right heart failure. On admission, the physical examination showed significant jugular venous distension, irregular cardiac rhythm with a blood pressure of 140/80 mmHg, and heart rate of 80 bpm. Her pulmonary auscultation was normal. The patient had voluminous ascites with significant lower limb edema. The electrocardiogram showed atrial fibrillation and right bundle-branch block. The laboratory tests were normal.

The echocardiogram showed a dilated and hypokinetic right ventricle, with a diastolic diameter of 3.8 cm, and left ventricular diastolic diameter and ejection fraction of 4.5 cm and 73%, respectively. The right atrium was enlarged. The pericardium was thickened with a mild effusion. The atrioventricular valvular filling was of the protodiastolic type with the E wave greater than the A wave. The clinical diagnostic hypothesis of severe restrictive cardiomyopathy with right predominance and significant tricuspid insufficiency was established.

The patient underwent cardiac catheterization, which showed coronary circulation without obstructive lesions, a normal left ventricle, and loss of the right ventricular intracavitary geography and amputation of the right ventricular trabecular region, suggesting the diagnosis of right ventricular endomyocardial fibrosis.

As the patient remained symptomatic with refractory ascites, requiring relief punctures every 15 days, surgical treatment was chosen, with resection of the right ventricular endomyocardial fibrosis and DeVega tricuspid valve annuloplasty.

The anatomicopathological examination of the surgical specimen confirmed the clinical and surgical impression, revealing "dense fibrosis of the endocardium, with a band containing neoformed vessels and calcified areas, an aspect compatible with endomyocardial fibrosis".

The patient evolved uneventfully in the immediate postoperative period, but, 3 months after the procedure, the huge ascites resumed. On August '97, the patient sought the emergency unit with an exteriorized abdominal hernia and a great flow of ascitic fluid. The patient remained refractory to clinical treatment, with ascites, congestive hepatomegaly and fatigue on minimum exertion, being then referred for heart transplantation assessment.

On that occasion, the echocardiogram evidenced a left ventricular ejection fraction of 69%, left atrium of 48 mm, and moderate mitral insufficiency. The right ventricle had hyperechogenic images, suggesting filling due to fibrosis and important tricuspid insufficiency on Doppler. On radionuclide ventriculography, the left ventricular ejection fraction was normal (55%), and the right ventricular ejection fraction was moderately depressed (36%). The cardiopulmonary test showed maximum oxygen consumption (VO_2) of 9.4 mL/kg/min. A new cardiac catheterization showed the following: no obstructive lesions in the coronary circulation; severe tricuspid insufficiency; mild mitral insufficiency; loss of the right ventricular intracavitary geography and amputation of the right ventricular trabecular region; an image suggestive of a fibrotic band in the left ventricular apex; and pericardial effusion. Cardiac output was 3.8 L/min, systemic vascular resistance was 1368 dyna/seg/cm^{-5}, pulmonary vascular resistance was 224 dyna/seg/cm^{-5}, and the cardiac index was 2.4 L/min/m^2.

The liver tests were normal and the abdominal ultrasonography evidenced signs of congestive hepatomegaly. Heart transplantation was indicated.

On May 15, 2000, the patient underwent bicaval orthotopic heart transplantation, whose postoperative complications were bleeding and pneumonia. She evolved satisfactorily, and received cyclophosphamide, azathioprine, and prednisone for rejection prophylaxis. The first endomyocardial biopsy (05/23) was zero degree, and the second biopsy (06/12) was 3A degree, and the patient was treated with prednisolone pulse for 3 consecutive days.

Currently, the patient's evolution is clinically satisfactory, with complete regression of the symptoms and ascites.

Discussion

Endomyocardial fibrosis is a rare disease in our country. Clinical treatment is indicated for less symptomatic patients, and the surgical treatment is indicated for more advanced stages (NYHA functional classes III and IV) [9]. The results are somewhat encouraging, mainly in patients with advanced disease and greater impairment of the right ventricle, such as in our patient.

The surgical results reported in the literature show an early mortality rate after surgery ranging from 4.6% to 18% [11,12] and a late mortality rate of 18% [11]. The maintenance of clinical symptomatology after surgery for resecting endomyocardial fibrosis in our patient may be attributed to the progressive characteristics of the disease, with continuation of the fibrotic myocardial infiltration and appearance of postoperative tricuspid insufficiency.

The Brazilian experience with 798 patients referred for heart transplantations comprises 407 patients with idiopathic dilated cardiomyopathy, 196 with ischemic heart disease, 117 with Chagas' disease, 29 with heart valvular disease, 14 with congenital heart diseases, 12 with peripartum heart disease, 7 with hypertrophic heart disease, 5 with restrictive heart disease, 4 with alcoholic heart disease, 1 with heart disease due to drug abuse, and 6 retransplantations [13]. No report on patients with endomyocardial fibrosis exists.

This case report stresses the need for careful clinical assessment of hepatic function in patients with right heart failure due to congestive hepatopathy secondary to heart disease, which may limit the receptor's survival and greatly increase the surgical risk. In these cases, simultaneous heart-liver transplantation, which is not routinely performed in our country, may be indicated.

The patient's clinical condition significantly improved after transplantation, but she still requires diuretics to control her abdominal volume.

In conclusion, heart transplantation may be an alternative for treating patients with endomyocardial fibrosis in advanced stages.

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