Improvements of Heart Failure by Cardiac Resynchronization Therapy in Cardiac Sarcoidosis

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In patients with cardiac sarcoidosis, progression of the disease may lead to end-stage heart failure that requires emergent mechanical homodynamic support or heart transplantation. This case was a 66-year-old female with cardiac sarcoidosis complicated with atrioventricular block and decompensated heart failure requiring a pacemaker implantation, but the treatment was in vain. Finally, cardiac resynchronization therapy (CRT) improved her heart function and clinical symptoms. In conclusion, CRT could improve the heart function in this case caused by this kind of infiltrative disease.

Key Words: Cardiac resynchronization therapy • Sarcoidosis

INTRODUCTION

Cardiac involvement in patients with sarcoidosis is being increasingly recognized and is associated with poor prognosis. Although environmental and genetic factors have been implicated in its pathogenesis, the etiology of cardiac sarcoidosis remains obscure. Clinical manifestations include advanced heart block, arrhythmias, and congestive heart failure. Progression of the disease in the heart may lead to complete atrioventricular (AV) block or ventricular arrhythmia that requires implantation of a permanent pacemaker or implantable cardioverter-defibrillator (ICD), or mechanical circulatory support as a bridge to heart transplantation. Even with a heart transplantation, recurrence can occur. This case was a 66-year-old female with cardiac sarcoidosis complicated with AV block and decompensated heart failure after pacemaker implantation. Cardiac resynchronization therapy (CRT) was chosen to improve her heart function and clinical symptoms.

CASE REPORT

A 66-year-old female patient with sarcoidosis was diagnosed by a bronchial biopsy in March of 1988. She underwent immunosuppressive therapy involving a total of four methotrexate pulse treatments in 1990. However, she began to suffer from heart failure in 1991. Echocardiography revealed poor left ventricular (LV) function (LV ejection fraction of 12%). Her clinical symptoms met the criteria of cardiac sarcoidosis. She was referred to our cardiovascular center for the treatment of her congestive heart failure. On December 15, 1997, she suffered from complete AV block, and a permanent pacemaker (VVIR mode) was implanted on December 18, 1997. The electrocardiography (ECG) revealed a right ventricular (RV) pacing rhythm and ventricular premature contractions (VPCs). Coronary artery angiography revealed normal coronary arteries in 2004. In the following years, she was often hospitalized because of
recurrent decompensated heart failure and VPCs with acute pulmonary edema (Figure 1, Panels A, B). A dys-synchrony study showed increase of the aortic pre-ejection delay (164 msec), interventricular dysynchrony (55 ms) and delayed activation of LV posterior wall (50 msec) by echocardiography.

The patient received optimal medical therapy including diuretics, angiotensin-converting enzyme inhibitors, spironolactone, beta-blockers, and steroids, but in vain. She received a CRT pacemaker (InSync III, Medtronic Inc., Minneapolis, Minnesota, USA) on May 10, 2006. Implantation was performed according to the standard technique for biventricular pacing, with the insertion of an LV lead (Attain Bipolar OTW, Model 4194, Medtronic Inc., Minneapolis, Minnesota, USA) for LV stimulation into the coronary sinus, in the posterior-lateral branch. A right atrial (RA) lead (CapSure SP Novus 5594, Medtronic Inc., Minneapolis, Minnesota, USA) was placed in the RA appendage, and a RV lead (Cap-Sure SP 4024, Medtronic Inc., Minneapolis, Minnesota, USA) was placed in the RV apex. The AV interval was optimized by the Ritter method one day after the implantation to achieve a maximal transmitral diastolic filling. After the CRT pacemaker implantation, the patient’s clinical condition improved dramatically. Her functional class improved from NYHA function class III-IV to I under the same medical therapy. The QRS duration shortened from 160 to 120 msec. One year later, her LV ejection fraction had improved from 12% to 23% and her LA (48 to 43 mm) and LV cardiac chamber sizes reduced (LV end diastolic diameter: 70 to 65 mm, LV end systolic diameter: 65 to 60 mm). The mitral regurgitation

![Figure 1.](image-url) (A) Twelve-lead ECG exhibits ventricular pacing with a wide QRS complex. The ventricular pacing rate was set at 60 ppm and QRS duration, 160 msec. (B) Chest radiography shows cardiomegaly with pulmonary edema after the implantation of a pacemaker (VVIR). (C) Twelve-lead ECG reveals regular atrial sensing and biventricular pacing with a narrower QRS complex (duration: 120 msec). (D) Chest radiography after the implantation of a CRT device reveals a smaller cardiac size than the previous one.
improved from severe to moderate. The thickness of the ventricular wall became prominent during the LV systolic phase. The thickness of the LV posterior wall even increased from 12 mm to 18 mm during the LV systolic phase (Figure 2, panels A and B). There was significant mechanical and electrical reverse remodeling.

DISCUSSION

Cardiac sarcoidosis may be diagnosed by an endomyocardial biopsy or extracardiac tissue proof. An excellent response can be achieved with steroid therapy in the early acute inflammatory stage. Progression of the disease may lead to end-stage heart failure that requires administration of the inotropic agents for intensive treatment or heart transplantation. In this case report of cardiac sarcoidosis associated with high degree AV block and LV dysfunction, the VVI(R) pacemaker mode was not adequate to improve her heart function, but CRT did improve it. The HOBI PACe trial clipped showed that in patients with LV dysfunction who needed permanent ventricular pacing support, CRT was superior to conventional RV pacing. According to the recent AHA/ACC/ESC 2008 guideline, patients with LVEF ≤35% with NYHA functional Class III or ambulatory Class IV symptoms who are receiving optimal recommended medical therapy and who have frequent dependence on ventricular pacing, CRT is reasonable (class IIa). In this case with cardiac sarcoidosis after the implantation of the CRT device, mechanical reverse remodeling occurred. The mechanism of cardiac function improvement may be due to restoration of AV sequential pacing, improvement of LV dyssynchrony and decrease of mitral regurgitation. This case report shows that CRT improved the clinical symptoms and cardiac function in a patient with cardiac sarcoidosis.

REFERENCES