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A Case of Loeffler's Endocarditis Associated with Churg-Strauss Syndrome

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Loeffler's endocarditis is generally caused by hypereosinophilic syndrome. It is a restrictive cardiomyopathy characterized with eosinophilia and eosionophilic penetration leading to the fibrous thickening of endocardium of both ventricles, apical obliteration and heart failure. We report a case of a 23-year-old male with Loeffler's endocarditis caused by Churg-Strauss syndrome. The echocardiogram showed that biventricular failure with large thrombus in left ventricle. His symptoms and typical echocardiographic findings markedly improved within 2 months after treatment for Churg-Strauss syndrome.

KEY WORDS: Loeffler's endocarditis · Churg-Strauss syndrome.

Introduction

Cardiac involvement is a major cause of morbidity and mortality in Churg-Strauss syndrome (CSS). It can be presented as cardiac arrest, myocardial infarction, vascular heart disease, congestive heart failure, pericardial effusion, and acute or chronic constrictive pericarditis.[1-6]

Loeffler's endomyocarditis is an inflammatory cardiac condition characterized by eosinophilic infiltration in the heart. However, thrombotic phase of Loeffler's endocarditis associated with heart failure in patients with CSS has been rarely reported.[7-9]

Case

A 23-year-old man was admitted to our hospital because of tingling sensation and swelling on the both calves, feet, and thighs. He also complained of dyspnea which developed 3 weeks ago. He had been suffering from bronchial asthma since he was a teenager.

On physical examinations, the patient did not have fever and showed rapid heart rate of 112 beats/min. Holosystolic murmur was heard at the apex. Decreased breath sounds were noted in both lower lung fields.

Laboratory examination revealed hemoglobin 12.2 g/dL, leukocyte count 11.5×10^9/L with 47.7% eosinophils (absolute eosionophil count 5.5×10^9/L), thrombocyte 316 $\times10^9$/L, aspertate amiotransferase 45 U/L, alanine aminotransferase 112 U/L. The X-ray of the thorax showed bilateral pleural effusion and subsegmental collapse and consolidation on right lower lung field (Fig. 1). Prominent soft tissues on the right tracheal and right hilar area were also noted. Electrocardiography showed a sinus rhythm with low voltages in standard leads, negative T-waves in leads I, II, III, and V2-6 (Fig. 2). Transthoracic echocardiography showed a mildly dilated left ventricle (LV) and left atrium, and biventricular systolic dysfunction (LV ejection fraction = 39%) with increased both ventricular myocardial echo density. Furthermore, diminished motion and echogenic thrombus-like mass (2.1×2.3 cm) were observed in the LV apex (Figs. 3A, B and C), with a moderate mitral valve insufficiency and small amount of pericardial effusion. Moderate tricuspid regurgitation was also shown on color Doppler image. Pulmonary artery systolic pressure was increased to 54 mmHg (peak tricuspid regurgitation velocity=3.5 m/s). Systolic mitral annular velocity (S') was reduced (4 cm/sec). Mitral inflow velocity and mitral annular velocity showed a marked restrictive LV filling; E, 105 cm/s, A, 44 cm/sec, E', 4 cm/s (Fig. 4A and B). Nerve conduction study suggested mononeuropathy multiplex. By all findings described above, he

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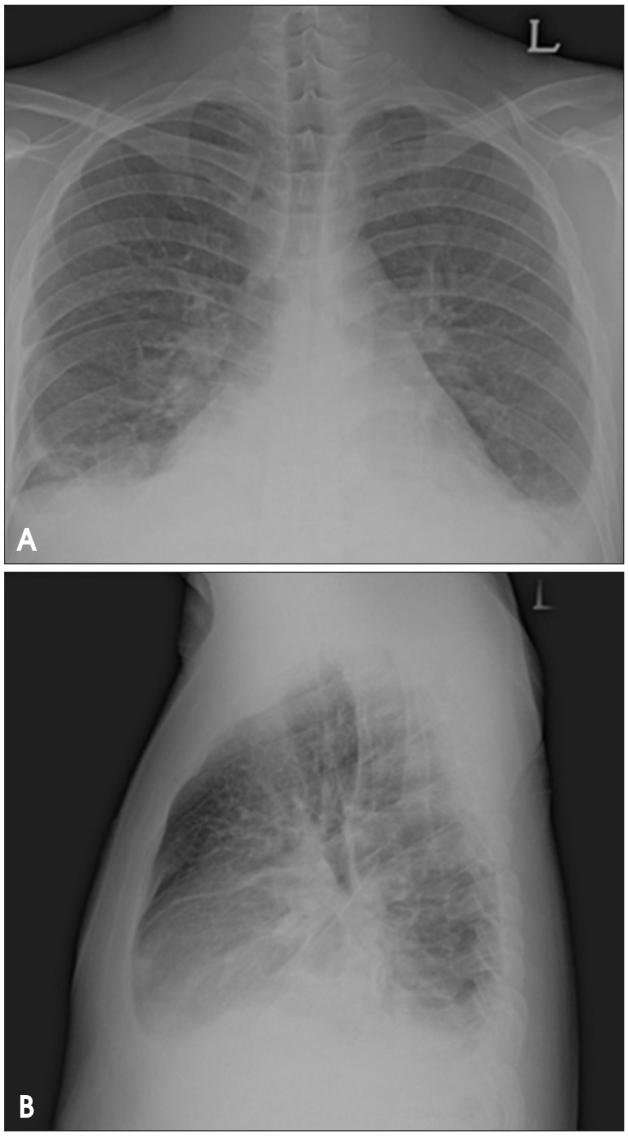

Fig. 1. Initial chest PA (A) and lateral (B) films show bilateral pleural effusion and subsegmental collapse and consolidation on right lower lung field.

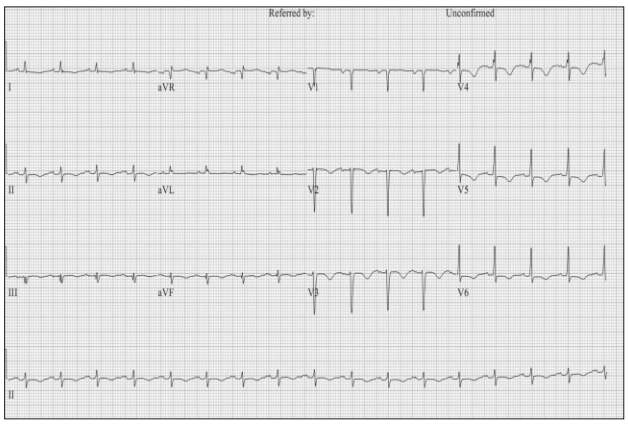

Fig. 2. Initial electrocardiogram shows tachycardia, low QRS-wave, and T-wave inversion in precordial and limb leads.

was diagnosed as CSS. Cardiac biopsy was not performed.

The patient was initially treated with methylprednisolone (1g/d for 1 week intravenously, then followed by 8 mg/d orally) and cyclophosphamide (200 mg/d). Anticoagulation therapy with warfarin was also performed. Medications for congestive heart failure were also started. The eosinophil count was decreased and clinical symptom was recovered rapidly after starting medications. Two months later, echocardiography showed markedly improved LV systolic function with an ejection fraction of 57%, and a decreased apical thrombus (Fig. 3D, E and F).

Discussion

We have demonstrated a case with Loeffler's endocarditis associated with CSS, which was improved after treatment for the underlying disease. The patient had 4 of the 6 conditions listed in the 1990 American College of Rheumatology classification criteria for distinguishing CSS from other vasculitides;[10] 1) a 10-year history of asthma, 2) peripheral eosinophilia (47.7%), 3) mononeuropathy multiplex, and 4) bilateral nonfixed pulmonary infiltration. In this case, we did not perform cardiac biopsy. Although endomyocardial biopsy is the gold standard for diagnosis, the biopsy can show false negative results and right ventricular biopsy sampling may miss the left-sided myocardial disease. Furthermore, biopsy on the left-sided chamber is not recommended because it may dislodge the mural thrombus, leading to systemic embolization.[11]

Loeffler's endomyocarditis generally shows 3-stage process: acute myocardial necrosis, microvascular fibrin deposition with or without thrombus formation, and endomyocardial fibrosis with resultant obliterative and restrictive cardiomyopathy.[7] The early necrotic stage is usually not recognized clinically. In this stage, a damage to the endocardium and infiltration of the myocardium with eosinophils and lymphocytes occur resulting in histopathologic evidence of myocardial necrosis, esosinophil degranulation and eosinophil microabscesses.[12] Echocardiography and angiography may not detect any abnormalities in this stage and endomyocardial biopsy is needed to make the diagnosis.[13] The second stage of heart disease involves formation of thrombi along the damaged endocardium of either or both ventricles and occasionally the atrium.[14][15] Finally, in the fibrotic stage, progressive scarring develops which may lead to entrapment of chordae tendineae with the development of mitral and/or tricuspid valve regurgitation and to endomyocardial fibrosis producing a restrictive cardiomyopathy.

Two-dimensional echocardiography is valuable in detecting intracardiac thrombi and the manifestations of fibrosis.[16-19] It was suggested that echocardiography should be performed in all patients with CSS to document the presence

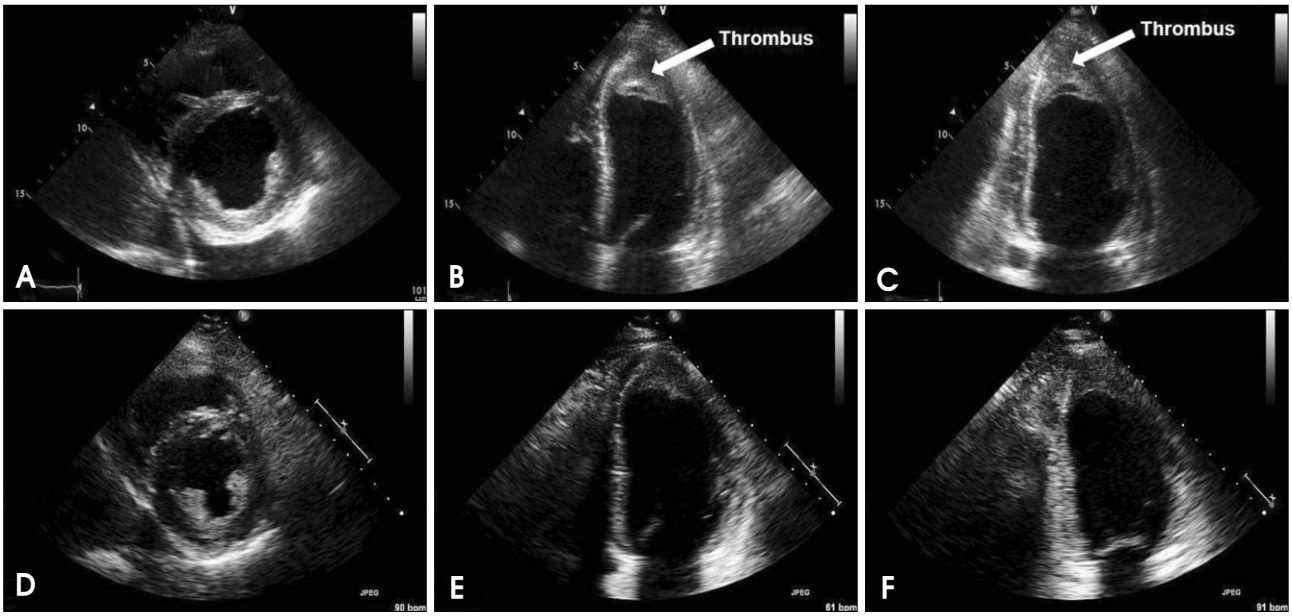

Fig. 3. Initial transthoracic echocardiography shows mildly enlarged left ventricle with depressed ejection fraction, and echogenic thrombus (2.1×2.3 cm) in the apex (arrows) (A, B and C). Left ventricular function is improved and size of the apical thrombus is decreased after 2 months of treatment for Churg-Strauss syndrome (D, E and F).

and extent of cardiac disease.[20] Echocardiography may facilitate early recognition of cardiac disease in CSS, such as valvular incompetence and myocardial fibrosis, which are usually subclinical, and thus improve the chances for effective reversibility.

The precise role of eosinophils as a causative factor in endomyocardial damage is unclear. An ultrastructural examination of cardiac tissue affected by eosinophilic endomyocarditis confirmed the close apposition of eosinophil to collagen bundles, a vital component of the myocardial connective tissue framework.[21] Evidence of eosinophil degranulation has been reported, and the release of eosinophil cationic protein, eosinophil major basic protein, eosinophil myeloperoxidase, or Charcot-Leyden crystal protein may be directly responsible for endomyocardial damage.[7] Eosinophil cationic protein and eosinophil major basic protein have been reported cytotoxic to endomyocardial cells.[22][23] The loss of endothelial integrity as a consequence of this may promote thrombosis and eventually fibrosis.[7]

In symptomatic patients, corticosteroid maintenance therapy is the first choice of treatment.[24] In steroid-resistant cases, myelosuppressive drugs like hydroxyurea or vinca-alkaloids can be tried.[25-27] Interferon-α also can be tried, which inhibits the degranulation of eosinophils.

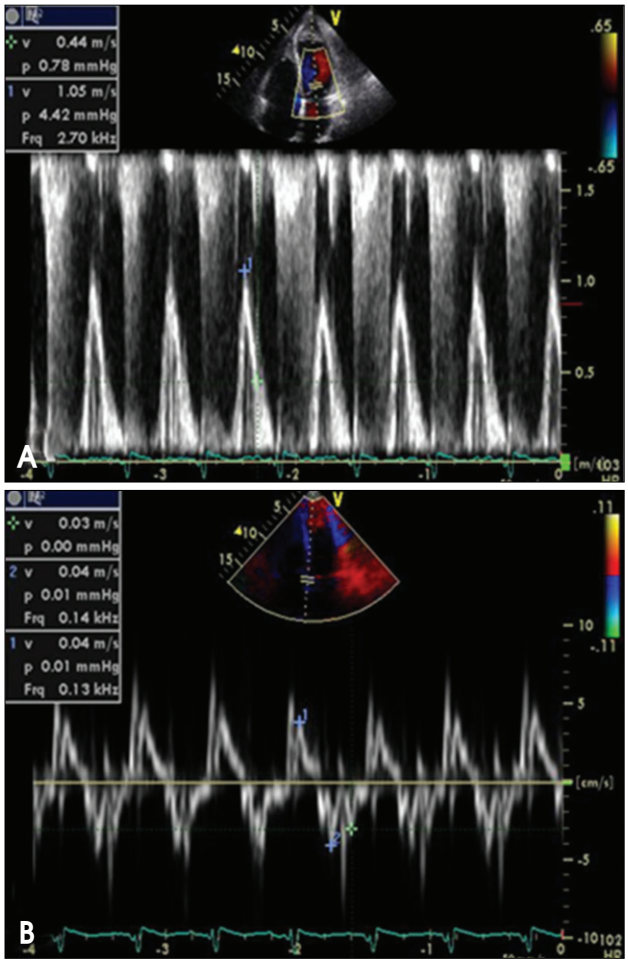

Fig. 4. Initial mitral inflow (A) and tissue Doppler image (B) show restrictive left ventricular filling pattern.

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