

Churg-Strauss Syndrome Presenting with Cardiac Mass and Right Ventricular Obstruction

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ABSTRACT

A 57-year-old woman presenting with asthma, hypereosinophilia, and generalized unspecific symptoms was diagnosed with Churg-Strauss syndrome. Echocardiography revealed a cardiac mass obliterating the right ventricle and severely impaired left ventricular function, which were caused by endomyocardial fibrosis. Cortisone and cyclophosphamide therapy resulted in amelioration of left ventricular function and significant size reduction of the right ventricular mass.

Introduction

Churg-Strauss syndrome is a granulomatous angiitis presenting with asthma and tissue and blood eosinophilia. Its characteristic clinical features are an infiltrative eosinophilic phase with similarities to Löeffler's syndrome, and finally a severe vasculitic phase. Cardiovascular manifestations include coronary vasculitis, acute and constrictive pericarditis, and myocardial fibrosis. Churg-Strauss associated cardiomyopathy is the most severe presentation, determining in most cases mortality and morbidity, while endomyocardial fibrosis has been rarely described.¹

Clinical Findings

We report the case of a 57-year-old white woman with a 5-month history of dry cough, 10 kg weight loss, increasing malaise, and exertional dyspnea. She had been hospitalized twice with the diagnosis of allergic asthma. At the time of the third admission we recognized a clinical constellation of asthma, moderate pericardial effusion, myocardial involvement (cardiac troponin I: 6.08 ng/mL), optical and radial neuropathy, and macular skin lesions. The laboratory tests showed inflammation (C-reactive protein: 110 mg/L, leukocytosis: 17 200/μL, erythrocyte sedimentation rate: 59 mm/1h), remarkable hypereosinophilia (3300/μL, 19%), and significant elevation of IgE levels. No cardiac arrhythmias occurred. Bone marrow biopsy revealed eosinophilic infiltration without malignant cells. Appropriate diagnostic measures for cardiac and extracardiac manifestations were

initiated and the diagnosis of Churg-Strauss syndrome² was confirmed.

Imaging

The striking finding at the first echocardiographic examination was a cardiac mass, almost completely obstructing the right ventricle (Figure 1). Moreover, there was severe impairment of left ventricular function with an ejection fraction of 27% and pericardial effusion without tamponade signs, but with fibrin strands. Using echocardiography alone, we were not able to distinguish the echogenic material obliterating the right ventricle from the adjacent myocardium. Cardiac magnetic resonance imaging (MRI) confirmed our echocardiographic findings and suggested that the echogenic mass within the right ventricle affecting the opening of the tricuspid valve might be a large thrombus. Furthermore, the contrast MRI exhibited circular, subendocardial late enhancement of the left and right ventricle, compatible with endomyocardial fibrosis.

For a more refined investigation of myocardial involvement tissue Doppler echocardiography with strain rate imaging was performed. We found normal longitudinal deformation (peak systolic strain -15% – 18% and peak strain rate -1.0 – 1.1 s^{-1}) without post-systolic shortening in the left ventricle and corresponding near-normal values in the right ventricle (peak systolic strain -17% and peak strain rate -1.7 s^{-1}). On the contrary, systolic and diastolic peak longitudinal myocardial velocities showed a significant reduction in both ventricles (peak systolic velocities:

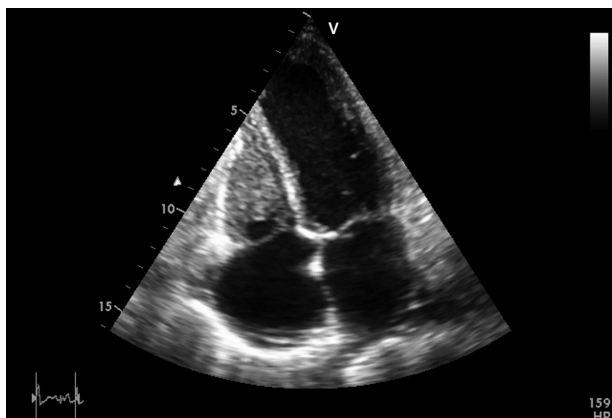


Figure 1. Initial echocardiographic examination: apical 4-chamber view showing a cardiac mass obliterating the cavity of the right ventricle.

3–4 cm/s in the left ventricle and 3 cm/s in the right ventricle; peak early diastolic velocities: $-5 - 6$ cm/s in the left ventricle and -4 cm/s in the right ventricle). This finding (normal deformation with severely reduced velocities) is highly suggestive for a diagnosis of endomyocardial fibrosis without intrinsic myocardial involvement.

Pathology

Endomyocardial left ventricular biopsy revealed fibrotic endocardial and myocardial changes with a relative fibrosis index of 3% (normal values below 0.5%). Myocyte necrosis or degeneration, as well as inflammatory reaction or eosinophilic infiltration of the myocardium were not observed by immunohistochemistry. The myocyte diameter was within normal range (18 μ m), excluding hypertrophic cardiomyopathy.

Clinical Course

Following cortisone pulse therapy, blood levels of eosinophilic granulocytes decreased significantly up to normal laboratory values within 10 days. After subsequent cyclophosphamide therapy the symptoms of heart failure ameliorated within the next month and the initially elevated levels of brain natriuretic peptide (9721 pg/mL) returned to near-normal values. However, serial echocardiographic evaluations showed persistent moderate systolic left ventricular dysfunction with ejection fraction of 40%. Interestingly, 4 weeks after initiating cortisone therapy, but without therapeutic anticoagulation, the right ventricular cardiac mass diminished considerably and the tricuspid inflow obstruction was completely resolved (Figure 2). Four months after diagnosis of Churg-Strauss syndrome and adequate therapy the patient still remains symptomatic, complaining of malaise, moderate dyspnea, and reduced exercise capacity.

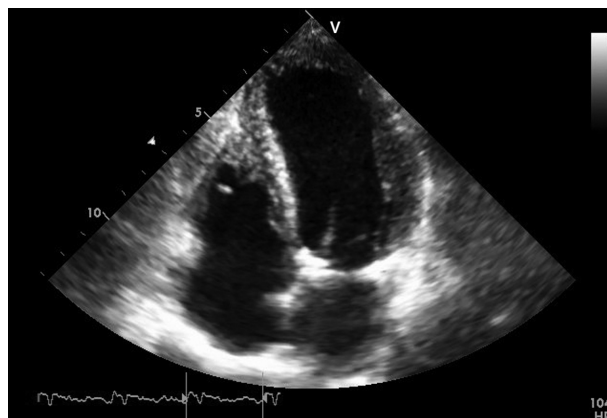


Figure 2. Follow-up echocardiography after 4 months: apical 4-chamber view demonstrating regression of the right ventricular mass.

Discussion

Patients with Churg-Strauss syndrome can present a variety of unspecific symptoms, especially at an early stage of this disease.³ Special attention is required to identify possible cardiac involvement, since cardiac manifestations frequently determine morbidity and mortality.

The particularity of this case is severe biventricular endomyocardial fibrosis with an intracardiac mass obliterating the right ventricle.⁴ The regression following steroid therapy without anticoagulation could be an indicator for its underlying pathophysiology: among direct eosinophilic degranulation, eosinophilic infiltration of the myocardium damages normal endothelial lining, thus exposing von Willebrand factor, tissue factor, and collagen. In turn, these factors are serving, through biochemical reactions, as stimuli for fibrin thrombus formation.⁵ However, in other cases with right ventricular obstruction a lack of response to steroids regarding the size of the cardiac mass has been reported.⁶ Since no right ventricular biopsy was performed, it remains unclear whether the origin of the cardiac mass was primarily eosinophilic or thrombotic.

Suspicion of myocarditis was raised initially by elevated troponin I and diffuse left ventricular hypokinesia, while hypereosinophilia was an indicator of Churg-Strauss associated cardiomyopathy.^{4,6} The clinically difficult differential diagnosis between myocarditis and endomyocardial fibrosis was correctly solved by echocardiography with tissue Doppler and strain rate imaging and further confirmed by MRI and histology findings.

Conclusion

This clinical presentation demonstrates the importance of echocardiography as a noninvasive and highly available technique in the primary assessment of patients with suspected Churg-Strauss syndrome. Characterization of regional myocardial function using tissue Doppler and

strain imaging is particularly useful to detect the pattern of cardiac involvement and to guide therapy. Besides the initial cardiac evaluation, a close echocardiographic follow-up is mandatory in the time-course of this vascular disease.

References

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