Medical Therapy Alone Produces Regression of Combined Aortic and Mitral Valve Involvement in Hypereosinophilic Syndrome

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A 36-year-old man presented with 1 month of fatigue and 5 days of gait disorder and agraphia. He had previously been diagnosed with hypereosinophilia. He was admitted to our hospital. On examination, his vital signs were stable, and he had a grade 2/6 systolic murmur at apex and a grade 2/6 diastolic murmur at the left sternal border. Clinical signs of heart failure were absent. Neurological examination revealed constructive apraxia, left-side impairment in skilled motor behavior, and right pyramidal tract sign. The ECG showed normal sinus rhythm and negative T waves at leads III and aVF. The chest radiograph yielded unremarkable results. White blood cell count was 23 000/mm³ with 66% eosinophils. There were no findings of secondary causes for hypereosinophilia such as neoplasm, allergies, autoimmune disease, or parasite infection. Multiple cerebral infarctions were found in his brain through magnetic resonance imaging (MRI). Transthoracic echocardiography (TTE) revealed mitral and aortic valve involvement but no thrombus in the ventricle (Figure 1 and Movie I in the online-only Data Supplement). Transesophageal echocardiography revealed aortic valve involvement with a mobile mass (sized 20×9 mm) attached to the right and left coronary cusps (Figure 2). Mitral valve involvement consisted of a mass joining the anterior to posterior mitral valve leaflets (Figure 3 and Movie II in the online-only Data Supplement). The 3-dimensional TTE (Artida, Toshiba Medical Systems Corp, Tochigi, Japan) enabled us to observe the mass from the point of view of the surgeon (Figure 4). In addition, color Doppler imaging showed mild regurgitation in both valves. In the cardiac MRI, delayed-enhancement sequence 10 minutes after infusion of 0.2 mmol Gd-diethylenetriamine pentaacetic acid per kilogram per square meter emphasized a hypersignal in the subendocardium around the anterior papillary muscle (Figure 5A through 5C). No thrombus in the ventricle was found on the MRI. The patient was diagnosed with idiopathic hypereosinophilic syndrome (IHES) with valve involvement and myocardial fibrosis. He was treated with anticoagulation and steroid therapy. The patient was discharged in stable clinical condition with a total white blood cell count of 11 400/mm³ with eosinophils 2%. At follow-up of the patient...
3 months later, TTE showed regression of masses on the aortic and mitral valves, although mitral and aortic regurgitation persisted mildly (Figures 1 and 3 and Movies III and IV in the online-only Data Supplement). Delayed-enhancement cardiac MRI showed a remaining hypersignal in the same anterior papillary muscle (Figure 5D through 5F).

IHES is a rare systemic disease defined by the combination of unexplained prolonged eosinophilia and evidence of organ involvement. Cardiac involvement is found in 54% to 73% of patients with IHES and is the major cause of mortality and morbidity.1 The atrioventricular valves are frequently involved, causing valvular regurgitation. Combined aortic and mitral valve involvement in IHES has been reported previously in only 2 case reports.2,3 These cases were treated with bivalvular replacement, with repeated replacement because of prosthetic valve dysfunction. Generally, additional difficulties with surgical approaches occur, and surgical therapy seems not to be of use in precluding a worsening prognosis. Two distinct features make our case worthy of reporting. First, cases with combined aortic and mitral valve involvement are extremely rare in IHES. Second, after medical therapy, there was echocardiographic evidence of regression of the previously seen masses on the aortic and mitral valves. To our knowledge, this is the first case in the literature in which anticoagulation and steroid therapy alone produced an observable regression in combined aortic and mitral valve involvement.

Disclosures
None.

References

Figure 2. The aortic valve mass (arrows) observed using transesophageal echocardiography from a longitudinal view (A, diastole; B, systole) and short-axis view in systole (C). LCC indicates left coronary cusp; RCC, right coronary cusp.

Figure 3. Comparison of mitral valve in parasternal short-axis views in diastole. The mass (arrows) on the mitral valve, which joined anterior to posterior leaflets on admission (A), achieved regression 3 months after initiation of medical therapy (B).
Figure 4. The mass (arrows) observed from the surgeon’s view by 3-dimensional TTE. AML indicates anterior mitral leaflet; PML, posterior mitral leaflet; LA, left atrium; and LV, left ventricle.

Figure 5. Delayed-enhancement images of cardiac MRI. MRI on admission revealed subendocardial delayed enhancement (arrows) around anterior papillary muscle in the short-axis (A), 4-chamber (B), and 2-chamber (C) views. MRI 3 months after initiation of medical therapy (D, E, F) showed small enlargement of the hypersignal area (arrows) indicating progressive endocardial fibrosis.
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