CASE REPORTS

Reversal of cardiac abnormalities in a young man with idiopathic hypereosinophilic syndrome using a tyrosine kinase inhibitor

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Abstract We report a case of idiopathic hypereosinophilic syndrome in a young man, who showed symptoms and electrocardiographic findings mimicking an acute coronary syndrome. Two-dimensional echocardiography, together with laboratory data, allowed us to make the diagnosis and to start a treatment with imatinib mesylate, a 2-phenylamonomopyrimidine-based tyrosine kinase inhibitor, which reversed the cardiac abnormalities.

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KEYWORDS
Echocardiography; Hypereosinophilic syndrome.

Case report

A 33-year-old man was admitted to the emergency department because of sudden onset of chest pain and shortness of breath at rest. On physical examination, he appeared diaphoretic with a blood pressure of 130/80 mmHg and a heart rate of 85 b/min. Cardiac auscultation revealed the presence of an olosystolic murmur and third heart sound at the apex, while crepitant rales were present in the mid and basal lung fields.

The electrocardiogram showed sinus rhythm with ST segment elevation in V1–V3 precordial leads and ST segment depression in lateral leads (Fig. 1). On the basis of these data, a diagnosis of acute coronary syndrome was made and the patient was transferred to our coronary care unit.

To better assess left ventricular function and to elucidate the cause of the olosystolic murmur, a transthoracic echocardiogram was performed with an ATL HDI-5000 CV digital system (Advanced Technology Laboratories, Bothell, U.S.A.) using a broad-band transducer with second harmonic capability (4–2 MHz). An apical long-axis view revealed normal left ventricular chamber size and function with obliteration of the apex by a large thrombus extending along the lateral wall to the posterior mitral leaflet that appeared, in a short-axis view, markedly limited in its motion.

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Color flow Doppler revealed a moderate mitral regurgitation with a jet impinging the lateral wall of the left atrium, while Pw-Doppler of mitral inflow showed a restrictive pattern with the E wave/A wave ratio >2 and a short deceleration time.

Laboratory values were in the normal range, except for white blood cell count, which was of 24,000/mm³ (70% eosinophils).

The patient was treated with diuretics, digitalis and heparin and his conditions markedly improved within a few hours. On the second day, the patient was transferred to the hematology division. Chest radiography and abdominal and pelvic ultrasound scans were performed to rule out a neoplastic disorder. Furthermore, the patient underwent a wide range of laboratory tests, including stool cultures, parasite serologies, IgE assessment, antinuclear antibodies and plasma cortisol evaluation. After such examinations and a bone marrow biopsy, a diagnosis of idiopathic hypereosinophilic syndrome was made. We decided to perform both a coronary angiography to confirm the absence of a significant coronary atherosclerosis and a percutaneous endomyocardial biopsy, but unfortunately, the patient refused to undergo an invasive assessment.

A treatment with imatinib mesylate (400 mg/die) and warfarin was started. After four months of this therapy, white blood cell count decreased to 7000/mm³ (4% eosinophils), the electrocardiogram showed normal sinus rhythm without ST segment modifications (Fig. 3), and no thrombus both in the apex and along the posterobasal left ventricular wall was present at the echocardiographic examination (Fig. 4).

Discussion

Idiopathic hypereosinophilic syndrome is a term labelling clinical illnesses characterized by blood hypereosinophilia and widespread infiltration of organs and tissues by mature eosinophils. The diagnosis is based on the criteria of Chusid et al.: sustained eosinophilia (more than 1500 eosinophils per mm³); the absence of other causes of eosinophilia, including parasitic infections and allergies; signs and symptoms of organ involvement. The involvement of the myocardium occurs in more than half of the patients, may be right and/or left sided, and usually indicates worse prognosis. Generally, the electrocardiogram shows non-specific ST segment and T wave abnormalities and arrhythmias, including atrial fibrillation and conduction defects.

A possible explanation for the unusual presentation in our young male relies on the fact that the eosinophil-mediated heart damage evolves through three stages: the acute necrotic stage, followed by the thrombotic stage which leads into the fibrotic stage. The acute necrotic stage, marked by an intense myocarditis with an associated arteritis, could explain the patient chest pain and the presence of Q-waves in V1–V3 precordial leads with concomitant ST segment elevation. In our patient, two-dimensional echocardiographic findings, i.e. apical obliteration by echogenic endomyocardial proliferations extending along the posterolateral wall and involving the posterior mitral leaflet, with normal global and regional function was the clue for the diagnosis.

Previously, Galiuto et al. reported a case of eosinophilic syndrome with an electrocardiographic presentation similar to a myocardial infarction,
Figure 2  (a) Apical long-axis view revealing a large thrombus extending along the lateral wall of the left ventricle; (b) the posterior mitral leaflet appeared markedly limited in its motion by the thrombus (parasternal short-axis view).
Figure 3  ECG performed after four months of therapy with imatinib mesylate.

Figure 4  Echocardiographic examination after four months of therapy, showing the absence of the thrombus.
but with the echocardiographic finding of left ventricular dysfunction in conjunction with apical asynergy and thinning.6

Recently, some cases of hypereosinophilic syndrome have been reported to respond to imatinib, a 2-phenylamnonopyrimidine-based tyrosine kinase inhibitor, which has been approved for the treatment of BRC—ABL positive chronic myeloid and acute lymphoblastic leukaemia.7–10 The clinical condition and the hematologic features of our patient markedly improved after one month of this therapy and after four months no electrocardiographic and echocardiographic abnormalities could be observed.

References