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CLINICAL CASE REPORT

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Atypical Presentation of Intracardiac Floating Thrombi in Hypereosinophilic Syndrome Complicated With Stroke and Systemic Embolization

A Case Report

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Abstract: Hypereosinophilic syndrome (HES) describes a disorder characterized by persistent peripheral blood eosinophilia with evidence of multiple target organs damage caused by eosinophilia. HES most commonly involves the heart, and cardiac involvement typically presents in the form of endomyocarditis or myocarditis with apical mural thrombus formation.

We present a case with atypical cardiac presentation with massive intracardiac fragile thrombi, causing peripheral emboli and strokes.

HES can present as floating thrombi with thin attachment to the left ventricle, and clinicians should also be vigilant of thromboembolic complications and initiate early therapy to prevent or reduce the potential complications of HES.

(*Medicine* 94(43):e1844)

Abbreviations: HES = hypereosinophilic syndrome, LV = left ventricule, MRI = magnetic resonance imaging.

INTRODUCTION

Hypereosinophilic syndrome (HES) describes a disorder characterized by persistent peripheral blood eosinophilia with evidence of multiple target organs damage caused by eosinophilia.[1] HES most commonly involves the heart, and cardiac involvement typically presents in the form of endomyocarditis or myocarditis with apical mural thrombus formation.[2] We present a case with atypical cardiac presentation with massive intracardiac fragile thrombi, causing peripheral emboli and strokes.

METHOD

This was a case report. The Institutional Review Board of the Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan, approved this study. Informed consent was obtained from family member of the patient.

CASE PRESENTATION

A 52-year-old male, with a history of well-controlled hypertension, was admitted to our hospital because of progressive dyspnea and orthopnea for 2 weeks. Orthopnea, general malaise, and generalized joint pain developed about 5 days later, along with some petechiae over his right palm. His white blood cell count was 33,200 mL with 51% eosinophil (absolute eosinophil count: 16,600 mm^3). The laboratory tests revealed elevated levels of CRP (11.97 mg/dl), troponin I (58.44 ng/m), and pro-BNP (20989 pg/mL), suggesting myocarditis with heart failure. Physical examination revealed lower limb edema, bilateral basal wheezing and diffuse erythematous papules and petechiae over his palms and abdomen (Fig. 1A–C). Neurologic examination showed right limb weakness with severity scale of 3 on a scale of 5 (scale: 0 = no movement; 3 = movement against gravity, but no resistance; 5 = normal muscle power). HES with myocarditis was highly suspected and was admitted for further evaluation.

Magnetic resonance imaging (MRI) of the brain revealed numerous foci of recent infarctions in bilateral cerebral and cerebellar regions (Fig. 1D). Transthoracic echocardiography showed mild left ventricle (LV) systolic dysfunction, global increased LV wall thickness (especially the posterior wall), and most importantly, several fragile and floating thrombi with thin attachments to the LV wall (Figure 2A and B, Videos 1–4 (Video 1—Echocardiography in left ventricular long axis view showed increased wall thickness in the posterior wall, suggesting myocardial fibrosis. Diffuse intracardiac thrombi with narrow attachment to the left ventricular endocardium were also seen. Video 2—Echocardiography in apical 4 chambers view showed diffuse intracardiac floating thrombi with narrow attachment to the left ventricular endocardium. Video 3—Echocardiography in apical 3 chambers view showed increased wall thickness over apical and middle segments of the posterior wall and floating thrombi. Video 4—Three-dimensional reconstruction of echocardiography in 4 chamber view

Editor: Alessandro Durante.
Received: April 2, 2015; revised: September 24, 2015; accepted: September 25, 2015.
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The authors have no conflicts of interest to disclose.

ISSN: 0025-7974
DOI: 10.1097/MD.0000000000001844

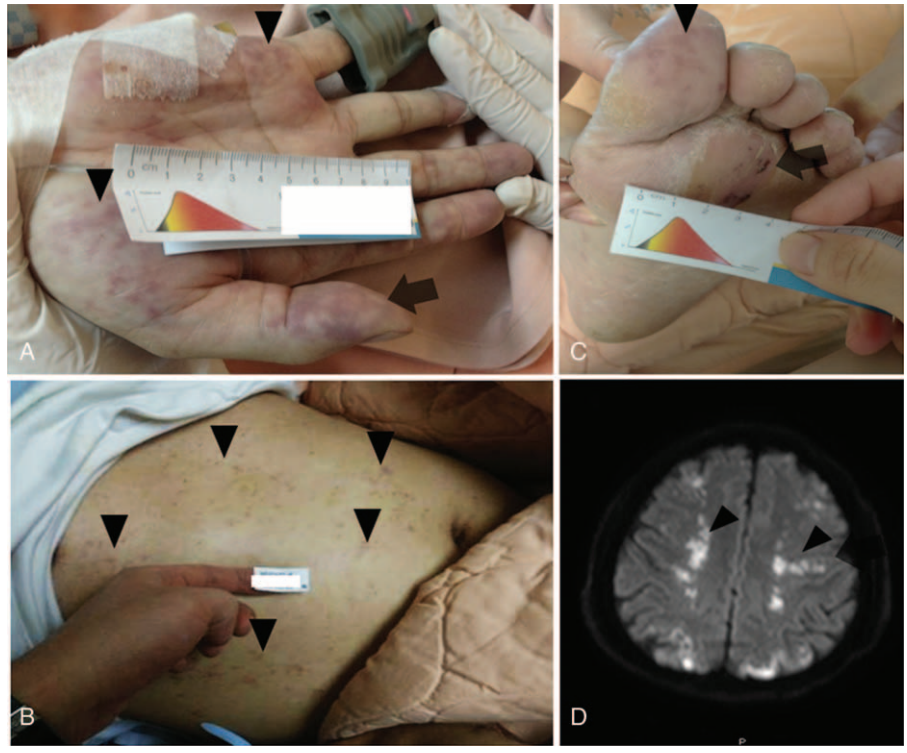

FIGURE 1. Hypereosinophilic syndrome (HES) complicated with diffuse brain and peripheral arterial embolizations. (A) Clinical photograph of the right palm showed erythematous papules and macules (arrowhead) over the right palm and livedo reticularis in the distal right thumb (arrow). (B) Clinical photograph of erythematous papules in the flank (arrowheads). (C) Clinical photograph of the right foot showed subcutaneous hemorrhage (arrow) over the left foot and purpura in the left toe (arrowhead). (D) Magnetic resonance imaging of the brain showed diffuse hyperintense signals from diffusion weighted image in bilateral cerebral hemisphere suggesting recent infarction (arrowheads). Similar embolic infarcts over bilateral cerebellum were also noted (not shown).

showed diffuse intracardiac thrombi, mainly in the left ventricular cavity.)). Analysis of other underlying autoimmune disorders and hypercoagulopathy all showed no significant abnormalities. HES with Loeffler endocarditis complicated with diffuse peripheral embolization and strokes was diagnosed, and medications with corticosteroid (methylprednisolone 1 mg/kg) and anticoagulants (heparin, at 2 times the normal value) were given. After 4 days of therapy, the eosinophila count

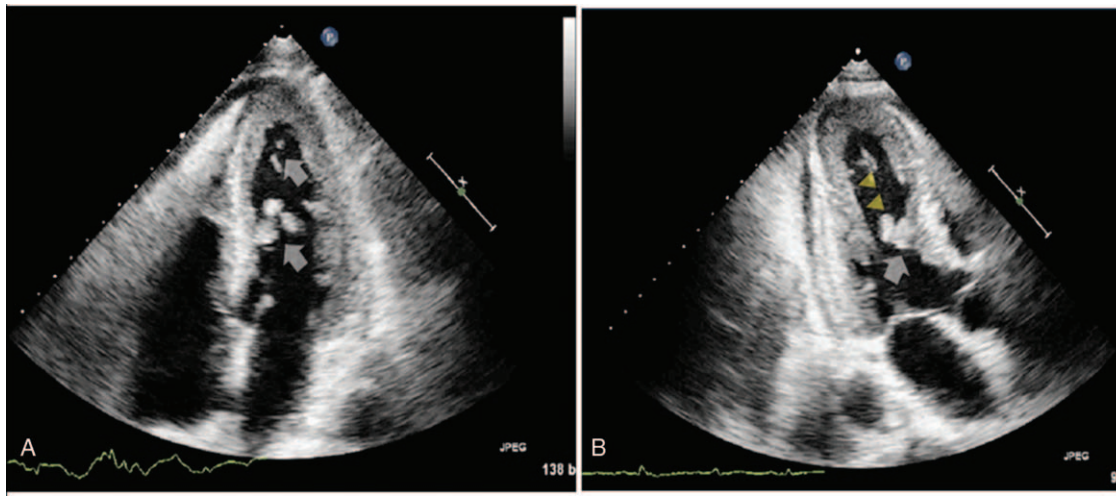

FIGURE 2. Transthoracic echocardiography showing endomyocardial fibrosis and diffuse intracardiac thrombus. (A) Four chamber view showed diffuse floating intracardiac thrombi (arrows). (B) Apical long axis view showed increased global wall thickness, especially apical and middle segment of the posterior wall (arrowheads) and floating thrombi (arrow).

returned to normal (absolute eosinophil count: 94 mm^3) and the symptoms of chest tightness and shortness of breath also improved. However, sudden onset of ventricular fibrillation developed on the fifth day, and despite our best efforts, he died on the same day.

DISCUSSION

HES is very rare and is characterized by the presence of hypereosinophilia (absolute eosinophil count >1500 cells/ mL on 2 examinations >1 month apart, and/or findings of tissue hypereosinophilia) with evidence of eosinophil-mediated target organ damage with all other potential causes of hypereosinophilia excluded.[1] In life-threatening end-organ damage, the diagnosis can be made immediately to avoid delays in therapy without waiting for tissue proof or imaging studies. Cardiac involvement, known as ''Loeffler endocarditis,'' is the most common manifestation (58%), and includes myocarditis, endocardial thrombi, endomyocardial fibrosis, myocardial ischemia, and finally restrictive cardiomyopathy, and it also responsible for the majority of deaths from HES (33–43%).[2,3]

The cardiac pathology of HES is divided into 3 stages: acute necrosis, thrombosis, and fibrosis.[4] The initial acute necrotic stage is characterized by eosinophilic infiltration and subsequent necrosis of the myocardium due to release of toxic cationic proteins from eosinophilic granules. Acute cardiac myonecrosis is followed by mural thrombosis formation, which is then replaced by fibrosis, leading to endomyocardial fibrosis, as in our case, and restrictive cardiomyopathy. Typically, the cardiac involvement presents in the form of endomyocarditis or myocarditis with apical mural thrombus formation.[2,5] Our case presented with atypical cardiac presentation with massive intracardiac fragile thrombus with mild thickening of LV wall. The fragile nature of these thrombi caused diffuse and severe peripheral emboli and strokes.

Reducing the eosinophilic count and regaining adequate cardiac function through medical or surgical approaches, including thrombectomy, endomyocardectomy, and even heart transplantation are possible options for patients with HES and cardiac involvement. The medical treatment for eosinophilia in patients with HES includes corticosteroid, hydroxyurea, immunosuppressant drugs, and tyrosine kinase inhibitor. Anticoagulants should be added for patient with evidence of intracardiac thrombi and thromboemboli with a significant improvement in 5-years survival of up to 80%.[3] However, despite improvement with therapy, the disease may progress rapidly with catastrophic outcomes as in our case.

CONCLUSION

In conclusion, clinicians should be aware that cardiac involvement is the most common manifestation of HES. It can present as floating thrombi with thin attachment to the LV, and clinicians should also be vigilant of thromboembolic complications and initiate early corticosteroid and anticoagulant therapy to prevent or reduce the potential complications of HES.

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