(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 5, Issue 4 , Page No: 501-505
July-August 2022

IJMSCR

Idiopathic Hypereosinophilic Syndrome With Cardiac Involvement: A Case Report

[1]Azin Alizadehasl, [2]Mahnaz Farajollahi, [3]Marjan Ghadri, [4]Mohammad Eslamijouybari, [5]Kamran Roudini, [6]Behnam darvishi,
M.D., [1]FACC, FASE,Professor & Echocardiologist, [6]Resident,
[1,2,3]Department of Cardiology, [4,5]Department of Hematology and Oncology,
[1,2,3]Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran
[6]Rasool Akram Hospital ,Iran University Of Medicine Tehran , Iran
[5]Cancer Research Center,Tehran University of Medical Sciences, Tehran, Iran
[4]Gastrointestinal Cancer Research Center, Mazandaran University of Medical Sciences, Sari, Iran

***Corresponding Author:**
Mahnaz Farajollahi
M.D, Department of Cardiology, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

We present the case of a 10-year-old boy who was diagnosed with idiopathic hypereosinophilic syndrome (IHES) and was under steroid therapy. He was referred to our center for more cardiac evaluations because of findings in transthoracic echocardiogram (TTE) that showed multiple masses in both ventricles during workups for constitutional symptoms .This case is presented to show the sooner the diagnosis is made with cardiac imaging and the specific treatments begin ,the better are chances to prevent the progression of the disease to later stages and we can decrease morbidity and mortality.

Keywords: idiopathic hypereosinophilic syndrome, biventricular thrombi, cardiac involvement

Introduction

Abbreviations: IHES: idiopathic hypereosinophilic syndrome. TTE: transthoracic echocardiogram. TEE : transesophageal echocardiogram. LV: Left Ventricle

History of presentation

A 10-year-old boy, who was diagnosed 6 months earlier with IHES and was under steroid therapy, was referred to our center due to multiple cardiac masses. The patient only suffered from weakness, fatigue, and mild bone pain that started a month earlier. No fever or dyspnea was reported. His drug history only included Prednisolone 30 mg once daily since he was diagnosed with IHES, and sometimes Acetaminophen if he had limb pain. His physical exam showed cushingoid face, no fever, normal and stable vital signs, and no lymphadenopathy. Heart auscultation revealed a normal S1 and S2 with a grade II/VI pansystolic murmur in the lower left sternal border. Lungs' auscultation was normal. Abdomen was soft and without organomegaly. Extremity examination showed no Osler's nodes, no Janeway lesions, no splinter hemorrhages.

Past medical history

Previous workups showed no history suggestive of allergy, drug reactions, malignancy, or parasitic infections. A bone marrow biopsy was performed, and it was normocellular with trilineage haematopoiesis and mild eosinophilia, but with no dysplastic or myeloproliferative features. Flow cytometry showed no evidence of any clonal lymphoproliferative disorder.

Differential diagnosis

The differential diagnosis of the cardiac mass was Vegetation and infections, primary or metastatic neoplasm, Loffler disease, clots or thrombus.

Investigations

In a complete blood count (CBC) test, the white-cell count was 110,000/µl, of which 90% was eosinophils. Renal function tests and liver function tests were normal. Blood culture samples were taken and empiric antibiotics were started. The initial TTE in the previous center showed multiple masses in both ventricles. An investigating TTE (Figure 1) was performed, followed by transesophageal echocardiogram (TEE) (Figure 2), to differentiate vegetation versus clot or other masses. And the results revealed a small left ventricle (LV) cavity size with preserved systolic function (LV ejection fraction: 50%) and thickening (13mm) of the LV wall (from the apex up to the mid part) with a large elongated mobile mass (12x50mm) attached to the LV apex mostly suggestive of a clot Normal right ventricle (RV) size with obliterated (thickness=12mm) apex and small (1.1x0.45cm) mobile mass in the RV apex mostly suggestive of a clot. Mild obstruction in the mid-cavitary of both ventricles due to masses (pressure gradient=35mmHg). Thickening of the posterobasal LV wall and diminished motion of the posterior Mitral valve leaflet resulting in moderate Mitral valve regurgitation. Due to the TEE results and negative blood cultures after a 7 day incubation period, infectious endocarditis was less likely, and the data were more consistent with multiple biventricular clots.

Management

Antibiotics were stopped and Intensive anticoagulation was initiated with low molecular weight heparin (LMWH) and oral vitamin K antagonist (VKA). After reaching the target International Normalized Ratio (INR) of 2.0-3.0.The patient was discharged with Warfarin and a continued prescription of Prednisolone (1 mg/kg/day).

Discussion

Idiopathic hypereosinophilic syndrome (IHES) consists of a group of disorders characterized by abnormal accumulation of eosinophils in the blood or peripheral tissues, independent of known secondary causes of eosinophilia such as parasitic infection [1]. Clinical manifestations of the condition are highly variable, ranging from asymptomatic eosinophilia to severe tissue damage and end-organ failure [2]. Although nearly any organ is prone to eosinophilia-associated damage, the heart is one of the most frequently targeted [3]. Eosinophilic myocarditis is a major cause of morbidity and mortality in patients; fatalities occur secondary to endomyocardial fibrosis and restrictive cardiomyopathy, and its incidence increases in the absence of therapy [4, 5]. The first mechanism of heart damage evolves through three stages, although these stages may be overlapping and not clearly sequential: (a) An acute necrotic stage in which the duration of illness has been short, with a mean of 5.5 weeks; (b) an intermediate phase characterized by thrombus formation along the damaged endocardium; and

(c) a fibrotic stage characterized by heart failure due to restrictive cardiomyopathy [6]. The disease is usually clinically silent in the acute necrotic phase and echocardiography can be normal during this stage. Several reports have shown that contrast-enhanced cardiac magnetic resonance imaging (MRI) reliably detects all stages and aspects of eosinophil-mediated heart damage, including the early stage of myocardial eosinophilic inflammation. The patients often develop cardiopulmonary symptoms and signs during the thrombotic and fibrotic stages. Another mechanism includes myocardial and pericardial damage from small vessel vasculitis. Echocardiography can evaluate myocardial function and MRI can demonstrate myocardial damage [7, 8]. Corticosteroid therapy during the acute stage may help control and prevent the evolution of myocardial fibrosis [9]. So earlier diagnosis of patients with IHES and the clinical and echocardiographic monitoring of heart disease, combined with use of cardiac medications, enables the cardiac sequelae of IHES to be managed more successfully and improves the longevity of IHES patients. A 1989 report of 40 HES patients from France, including 17 with the more serious features of a myelodysplastic syndrome, noted an 80% survival at 5 years and a 42% survival at 10 and 15 years [10]. So the most important point is to start appropriate drugs in the early stage of cardiac involvement before reaching the thrombotic and fibrotic stages, to reduce cardiac mortality and morbidities.

Follow-up

Volume 5, Issue 4; July-August 2022; Page No 501-505

The INR target were checked regularly, and a follow-up TTE that was performed 3 months later showed a reduction in the size of clots, but the corticosteroids couldn't be tapered because of an increase in eosinophils due to prednisolone dose reduction.

Conclusions

We described the case of an IHES patient who presented with multiple large biventricular clots that showed the cardiac involvement is at least in stage 2. IHES is an uncommon heterogeneous disease syndrome that requires a high clinical suspicion index so as to enable early diagnosis. Cardiac involvement of HES is associated with unfavorable clinical outcomes and therefore timely medical treatment is crucial to prevent the progression of cardiac morbidities. Recent advances in diagnostic imaging and a better understanding of IHES pathogenesis have resulted in non-invasive diagnostic opportunities and novel targeted therapies, respectively.

Learning objectives

1. Cardiac involvement is a major cause of mortality and morbidity in IHES and potentially lethal cardiac involvement increases in the absence of therapy so time is the most important point about diagnosing it and every effort should be done to detect it before organ damage and fibrotic stage.
2. Non-invasive modalities like cardiac MR and TTE could detect the involvement before clinical manifestations and high index of suspicion should be presumed as IHES is diagnosed.

References

1. Chusid MJ, Dale DC, West BC, Wolff SM. The hypereosinophilic syndrome: analysis of fourteen cases with review of the literature. Medicine (Baltimore) 1975;54:1–27.
2. Ogbogu PU, Bochner BS, Butterfield JH, Gleich GJ, Huss-Marp J, Kahn JE, Leiferman KM, Nutman TB, Pfab F, Ring J, Rothenberg ME, Roufosse F, Sajous MH, Sheikh J, Simon D, Simon HU, Stein ML, Wardlaw A, Weller PF, Klion AD. Hypereosinophilic syndrome: a multicenter, retrospective analysis of clinical characteristics and response to therapy. J Allergy Clin Immunol. 2009;124:1319–1325.e3.
3. Roufosse F, Cogan E, Goldman M (2004) Recent advances in pathogenesis and management of hypereosinophilic syndromes. Allergy 59:673–689.
4. Parrillo JE, Borer JS, Henry WL, Wolff SM, Fauci AS (1979) The cardiovascular manifestations of the hypereosinophilic syndrome. Prospective study of 26 patients, with review of the literature. Am J Med 67(4):572–582.
5. Legrand F, Renneville A, Macintyre E, et al. The Spectrum of FIP1L1-PDGFRA- Associated Chronic Eosinophilic Leukemia:New Insights Based on a Survey of 44 Cases. Medicine (Baltimore) 2013.
6. Weller PF, Bubley GJ. The idiopathic hypereosinophilic syndrome. Blood 1994; 83:2759.
7. Debl K, Djavidani B, Buchner S, et al. Time course of eosinophilic myocarditis visualized by CMR. J Cardiovasc Magn Reson 2008; 10:21.
8. Azin Alizadehasl ,Anita Sadeghpour,Chapter 5 - Echocardiography,Editor(s): Majid Maleki, Azin Alizadehasl, Majid Haghjoo,Practical Cardiology (Second Edition),Elsevier,2022,Pages 67-110.
9. Uetsuka Y, Kasahara S, Tanaka N, Sekiguchi M, Hiroe M, Yu ZX, Hosoda S.Hemodynamic and scintigraphic improvement after steroid therapy in a case with acute eosinophilic heart disease Heart Vessels Suppl, 5 (1990), p. 8.
10. Lefebvre C, Bletry O, Degoulet P, Guillevin L, Bentata-Pessayre M, Le Thi HD, Godeau P Facteurs pronostiques du syndrome hyperéosinophilique. Étude de 40 observations ,Ann Med Interne (Paris), 140 (1989), p. 253

Figures

Volume 5, Issue 4; July-August 2022; Page No 501-505

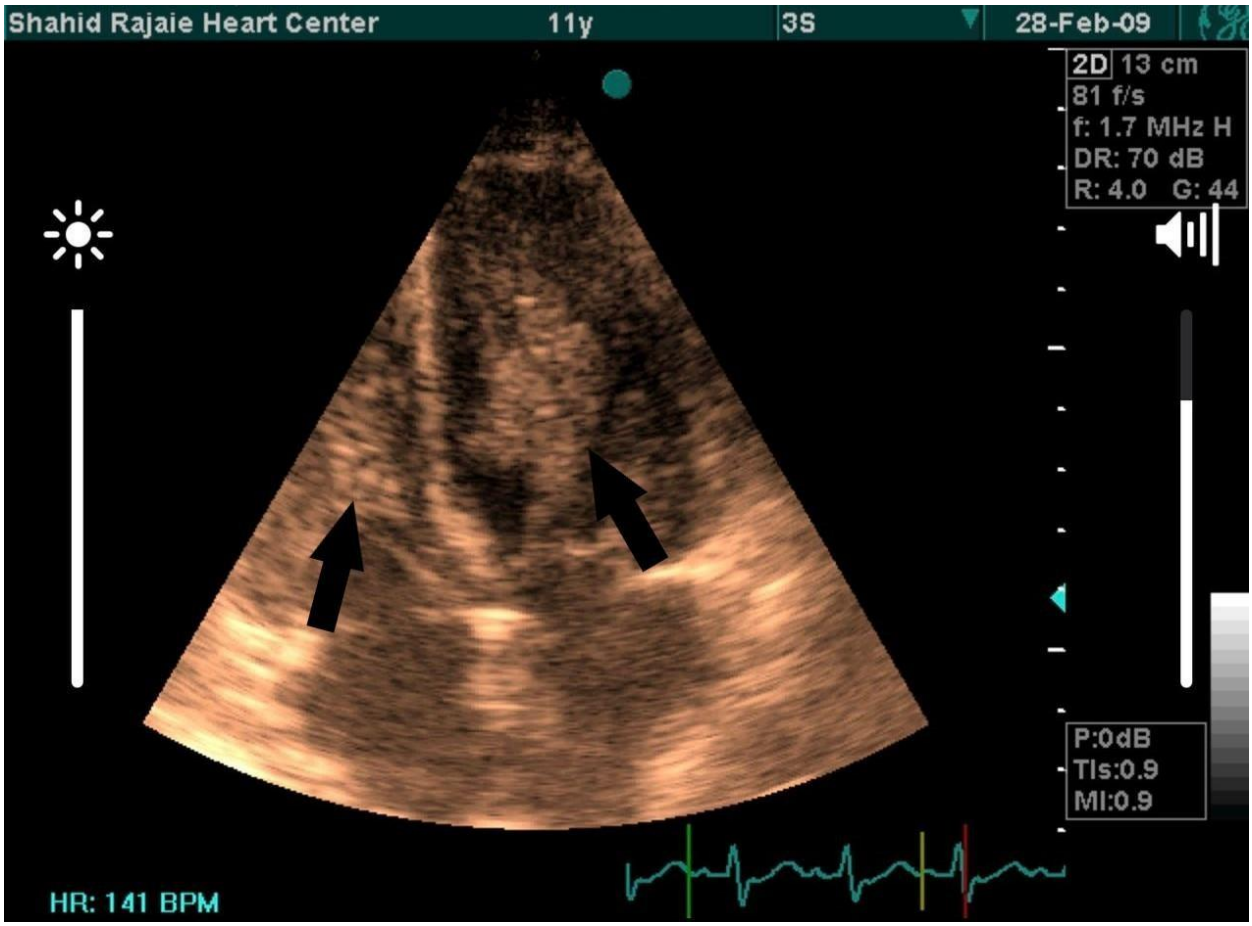

Figure 1 Transthoracic echocardiogram apical four-chamber view showing two large masses (arrows) in right and left ventricle.

Volume 5, Issue 4; July-August 2022; Page No 501-505

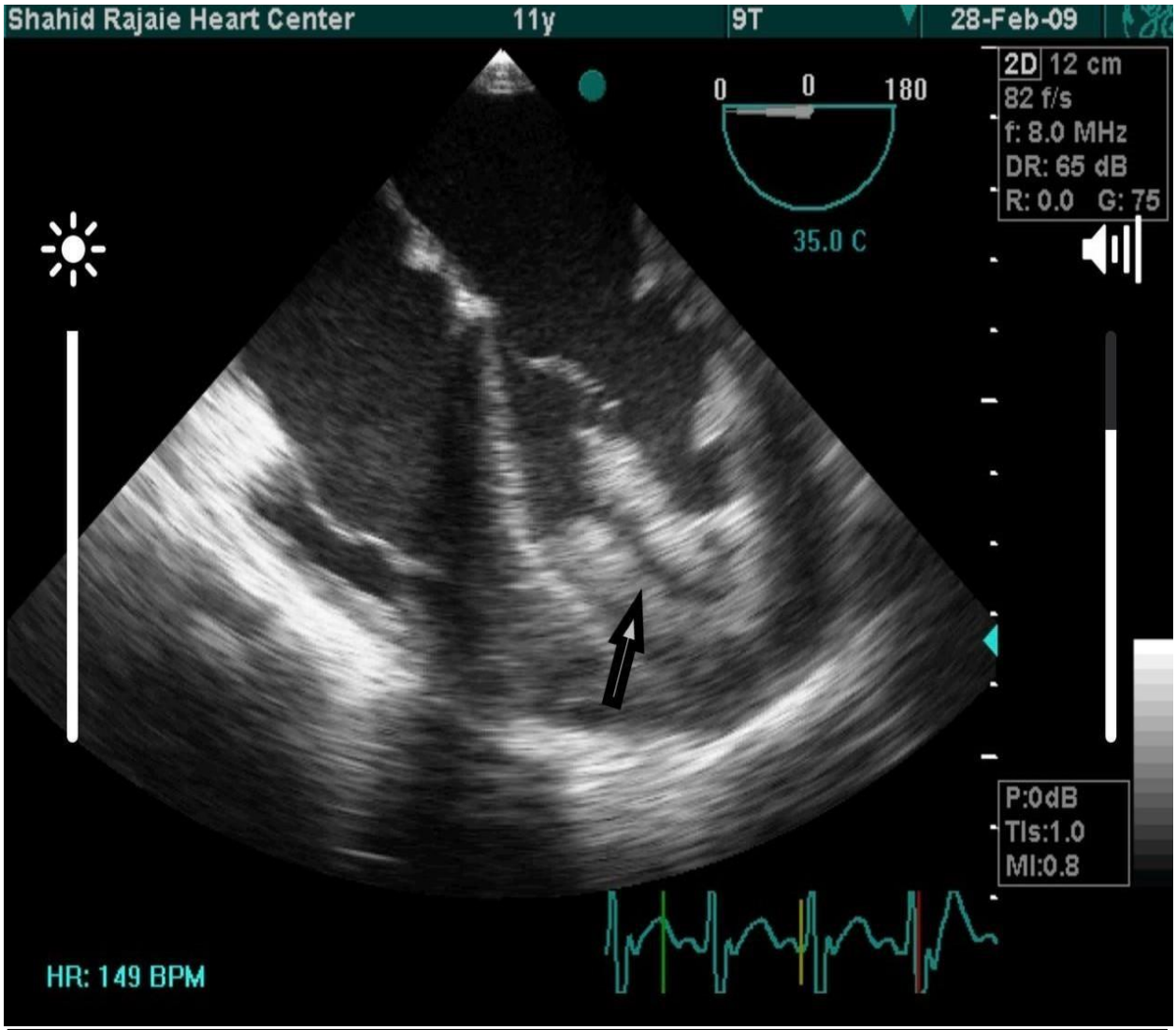

Figure 2 Four cavities view with transesophageal echocardiography showing a massive thrombus in the left ventricle (arrow)

Volume 5, Issue 4; July-August 2022; Page No 501-505