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Hypereosinophilic Syndrome Presented by Hear Failure Preserved Ejection Fraction: A Case Report of The Importance of Multimodality Cardiac Imaging in Loeffler Endocarditis

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1. Abstract

Loeffler endocarditis is a rare restrictive cardiomyopathy caused by hypereosinophilic syndrome, characterized by hypereosinophilia and fibrous thickening of the endocardium causing progressive onset of heart failure, thrombosis and valvulopathy. Herein we present a 47-year-old woman presented by rapidly progressive heart failure symptoms with preserved ejection fraction and atrioventricular valvulopathy. In the workup, endomyoardial thickening, atrioventricular valves regurgitations and restrictive cardiomyopathy by echocardiography. What is more, hyper intake of gadolinium by cardiac magnetic resonance imaging (CMR) associated with hyper eosinophilia in the blood investigation. After exclusion of other cause of hypereosinophilia, diagnosis of endomyoardial fibrosis (Loeffler endocarditis) in the context of a hypereosinophilic syndrome (HES) is therefore retained. In occlusion; echocardiography and CMR is a highly useful non-invasive modalities for the diagnosis of cardiac disease in HES patients.

2. Introduction

The notion of hypereosinophilic syndrome (HES) was introduced firstly in 1968 by Hardy and Anderson, defined by persistent blood eosinophilia without obvious cause, multiorgan involvement and fatal outcome [1]. Later in 1975, the diagnostic criteria of primary or idiopathic HES were stated by Chusid et al. and it is including: persistent blood absolute eosinophil count over 1500/mm3 for duration of more than 6 months, with evidence of tissue and organ damage, without any identifiable cause of eosinophilia [2] Prevalence and overall incidence have not been well defined. HES may affect the heart in approximately in up to 50% of HES [3]. The modern definition of HES because of the availability of more sophisticated equipments to rapidly evaluate eosinophilia and the need to start the treatment to minimize organ damage has change the diagnostic criteria were the eosinophilia persists for more than 6 months is less consistently embraced today; instead the duration of one or more month is recommended. [4, 5]. Loeffler endocarditis is an uncommon, but known complication of hypereosinophilic syndrome (HES). It is a relatively rare entity, and remains poorly understood, it is characterized by endomyoardial fibrosis (EMF), and it is firstly described in 1936 by Loeffler, who termed it "fibroplastic parietal endocarditis with blood eosinophilia" It is a relatively rare and an uncommon cause of restrictive cardiomyopathy (RCM) [6,7].

Herein we describe a case of a 47-year-old woman with Loffler's endocarditis presented with heart failure and atrioventricular valves regurgitation, this case demonstrates the use of echocardiography and cardiac magnetic resonance imaging (CMR) as a noninvasive and reliable tool to diagnose Loffler's endocarditis.

3. Case Presentation

A 47-year-old woman was admitted with severe shortness of breath New York Heart Association class 4 (NYHA V), typical angina pectoris, and elevated blood pressure 2 months before admission with progressive increasing in the symptoms. The physical examination at the time of admission the patient's blood pressure 115/75

mmHg, Spo2 = 91%, pulse rate = 105 beats per minute, mitral and tricuspid holosystolic murmur (S3 +, S4 +), crepitation in both lungs, and pedal edema (++). The electrocardiogram (ECG) shows sinus rhythm, 0.5 mm depression of ST segment in II, III, avF, and V3-V6 conduction, negative T tooth (Figure 1).

At the time of admission her Echocardiography shows left ventricular ejection fraction 50%, mass filling 2/3 of the left ventricular cavity, severe tricuspid regurgitation, mild mitral regurgiation, SPAP = 65 mm Hg, TAPSE = 18 mm, the transmitral flow E/A ratio 1.1/0.4, deceleration time (DT), E/e' ratio, no special comment on the valves (Figure 2). The CMR reveled endomiocardial fibrosis further comment on the gadolinium uptake and on the valve and others (Figure 3, 4). Furthermore, CT scan revealed bilateral pleural effusion. Laboratory tests revealed multiple elevated eosinophil (58%, 13,360 mm3). The patient underwent pleural puncture twice at different times with plural fluid analysis showed. A multidisciplinary team of cardiologist, hematologist, rheumatologist and pulmonologist was discussing the case, and the professional diagnosis of loefler's endocarditis induced by HES has been made. Steroid therapy please specify (type, dose rote and duration) concomitant with anticoagulant therapy (warfarin) dose and her base line INR and her INR on warfarin and for how long she received the warfarin. 1 and 3 month control laboratory (Eosinophilia 0%) and EchoCG examinations showed clinical + dynamics in the patient (LVEF = 50%, left interventricular mass decrease, mild mitral regurgitaion, minimal tricuspid regurgitation, SPAP = 25 mm Hg, pseudo normal diastolic dysfunction (Figure 5, 6).

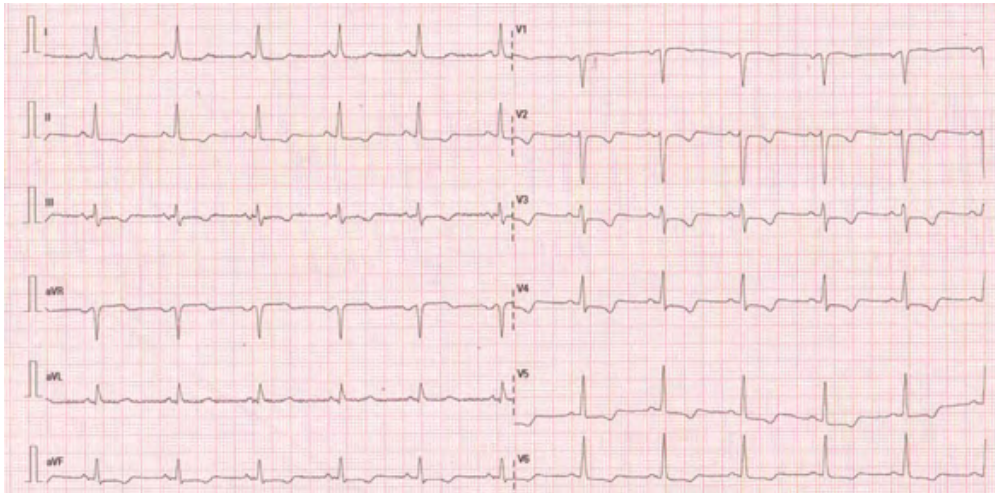

Figure 1: ECG.

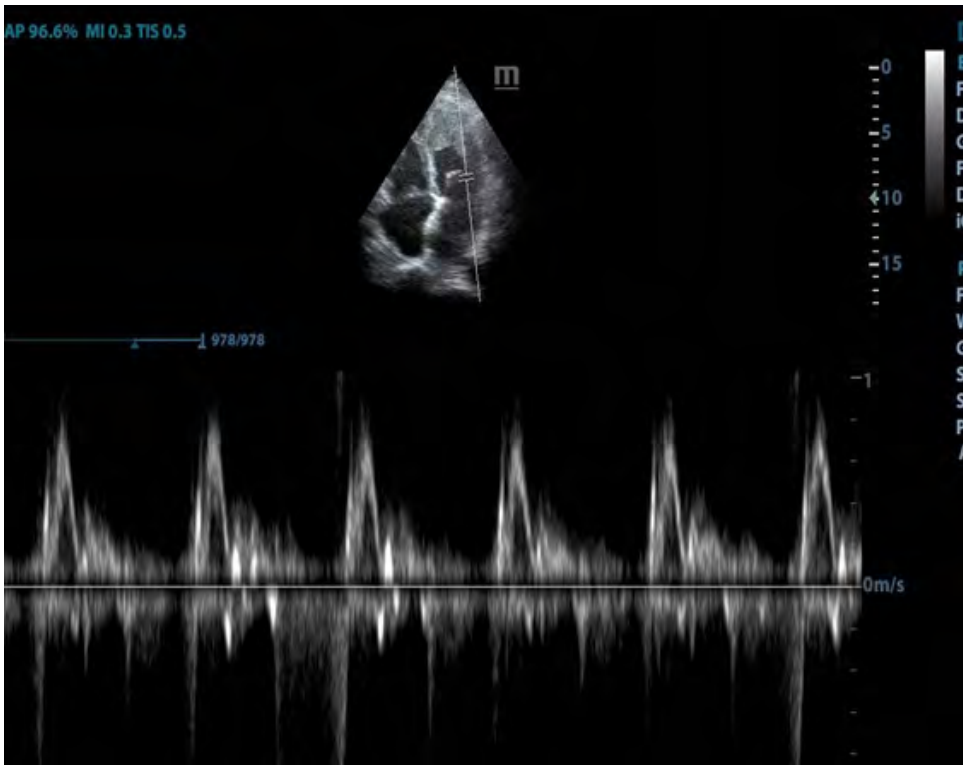

Figure 2: Transmitral flow.

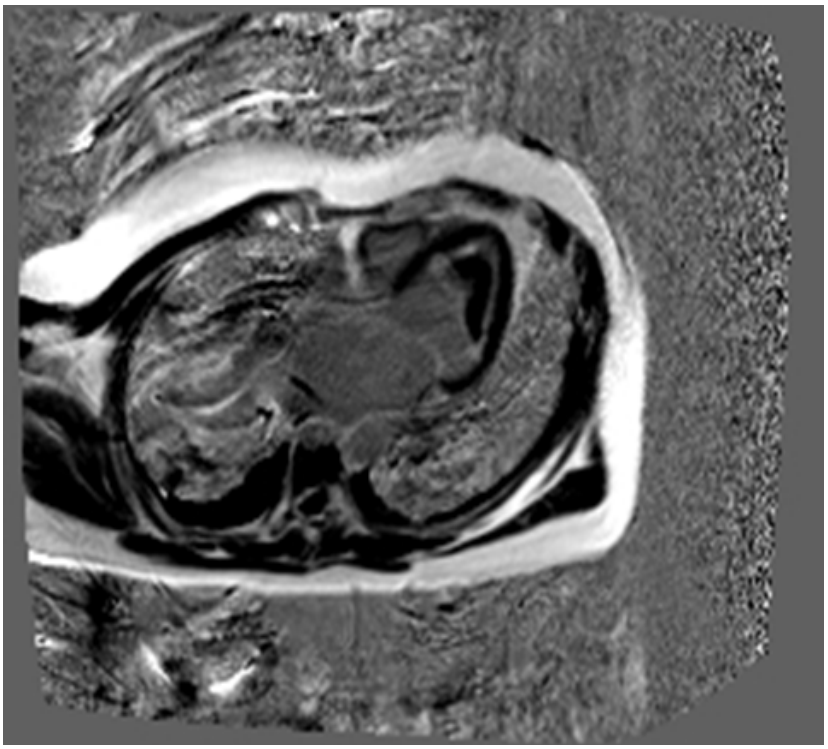

Figure 3: CMR 4CH view LGE.

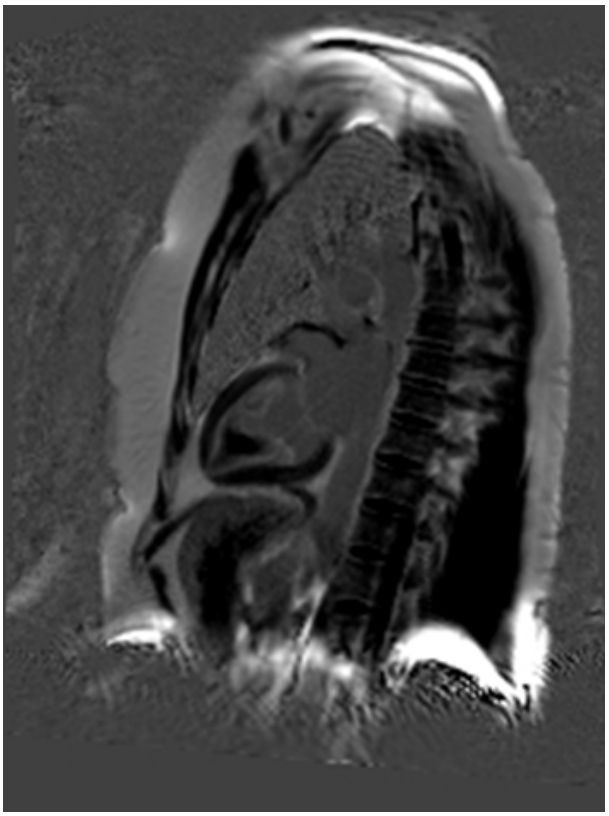

Figure 3: CMR 2CH view LGE.

4. Discussion

Historically the HES is defined as hypereiosenophilia of more than 1500mm3 for more than 6 month with endorgan damage, however; the modern definition of HES, because of the availability of more sophisticated techniques and the need to receive expedited treatment to minimize organ damage, has change the diagnostic criteria were the eosinophilia persist for more than 6 months is less consistently embraced today instead the duration of one or more month is recommended [4, 5]. The International Working Group on Eosinophil Disorders (ICOG-EO) classification of eosinophilic disorders in 2011 classify into following terms: hereditary (familial) HE variant; primary (clonal/neoplastic) HE variant; secondary (reactive) HE; and HE of undetermined significance/idiopathic variant [8]. Cardiac involvement could be present approximately in up to 50% of HES [3] and the most characteristic cardiac abnormality in HES is EMF (Loeffler endocarditis). It is a relatively rare conditions and an uncommon cause of restrictive cardiomyopathy (RCM) characterized by fibrosis that obliterates the ventricles with EMF as the ultimate form of eosinophilic cardiac disease [6, 7, 9] The natural cause of cardiac pathology in HES chronologically divided into three stages: eosinophilic infiltration, thrombosis and fibrosis [10, 11]. In a literature review of 26 case report of Loeffler endocarditis with hypereosenophic syndrome as the culprit cause, the most common cardiac structure affected was the mitral valve (65%), followed by the tricuspid valve (42%), left ventricle (23%), right ventricle (8%), right atrium (4%) and interventricular septum (4%) [12] Main echocardiographic signs are elevated filling pressure, progressive endomyocardial thickening, valve regurgitation, and possible intracardiac thrombus formation, Echocardiography is the mainstay of diagnostic imaging and surveillance for Loeffler endocarditis caused by HES. An national institution of health (NIH) study of 22 HES patients who had echocardiographs showed that 68% had left ventricular wall thickening, 37% had increased left atrial transverse dimension and 27% had an increase in right ventricular transverse dimension.[3] A Mayo clinic study consisting of 55 patients with hypereosinophilic syndromes and echocardiograms showed that 12% had endocardial thickening, 24% had left ventricular apical thrombus, 20% had right ventricular apical thrombus, 20% had posterior mitral leaflet involvement, 10% had tricuspid involvement, 16% had hyperdynamic LV, 10% had LV hypertrophy, 14% had LV dilation and 18% had pericardial effusion[10]. Echocardiographic series reported mitral regurgitation in 43% of HES Patients, the accumulation of thrombofibrotic material between the mural endocardium of the LV free wall and the ventricular side of the posterior mitral leaflet restricted posterior mitral leaflet motion [13] The cardiacmagnetic resonance imaging (CMR) findings are endomyocardial involvement, particularly late gadolinium enhancement (LGE) that shows extensive eosinophilic infiltrates that cause endomyocardial fibrosis [14-17] the late gadolinium hyper-enhancement in the apical region demonstrates fibrosis occupation and provides a definitive diagnosis of endomyocardial fibrosis, and ruling out the thrombotic component. ECG changes in HES patients, according to the Parrillo study are: LA enlargement, LV hypertrophy, ventricular premature complexes, poor R-wave progression, non-specific ST-T changes and first-degree heart block [3]. However, the role of ECG in diagnosis of HES is limited as it is provide only evidence of cardiac pathology no a specific evidence of abnormalities associated with HES [13]. Endomyoardial biopsy (EMB) is the gold stander diagnostic tool of the Loeffler endocarditis and the findings include fibrotic thickening of the endocardium, mural thrombosis and fibrinoid changes, thrombosis and inflammation of small intramural coronary vessels and, sometimes, eosinophil infiltration into the myocardium and sometimes endocardium [13]. Initiation of therapy is crucial. High-dose systemic glucocorticoids (prednisone 0.5–1mg/kg/day) are normally first-line agent for treatment of most patients who present with life-threatening or potentially disabling manifestations. Hydroxyurea is the most commonly used second-line [18].

5. Conclusion

Echocardiography and CMR is highly useful non-invasive modalities for the diagnosis of cardiac disease in HES patients.

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