International Journal of Clinical Images and Medical Reviews

ISSN 2771-6309

Case Report

Eosinophilic Myocarditis presenting with progressive Cardiac Cachexia

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Abstract

Background: Eosinophilic myocarditis, cardiac manifestation of hypereosinophilic syndrome (HES), occurs in HES patients frequently and represents a major cause of their morbidity and mortality. We present a case of a patient with advanced stage of eosinophilic myocarditis manifesting with progressive cardiac cachexia, despite intensive treatment.

Case presentation: A 66-female patient underwent comprehensive cancer screening due to significant weight reduction: patient's initial weight of 79 kg was gradually reduced to 49 kg during past 3 years, what represents a total weight loss of 38%. Cancer-screening showed negative results except for echocardiography, which revealed a large right ventricular mass imitating malignant cardiac tumor. Biopsy of the intracardiac mass was performed. However, the procedure was complicated with right ventricular perforation causing cardiac tamponade, successfully resolved by immediate pericardiocentesis. Subsequent echocardiography revealed an echodense mass also in left ventricular apex. Laboratory findings with mildly elevated eosinophil count and presence of formations in both ventricular apexes arose suspicion of non-malignant disease origin, specifically eosinophilic myocarditis. Cardiac magnetic resonance (CMR) imaging confirmed distinctive signs of eosinophilic myocarditis of both ventricles, together with presence of intracardiac thrombi located in right and left ventricular apexes, right atrium and right atrial auricle. Histologic findings from biopsy definitely ruled out formerly presumed diagnosis of cardiac tumor, since no signs of malignant proliferation was present. Evaluation of hemopericardium revealed significant concentration of eosinophils, without cytogenetic or molecular abnormalities, characteristic for idiopathic hypereosinophilic syndrome.

Conclusion: Presence of intracardiac formations in both ventricular apexes should raise suspicion of eosinophilic myocarditis, even in absence of significant peripheral hypereosinophilia. Our case highlights the importance of CMR imaging in diagnostic process of eosinophilic myocarditis, since it represents a noninvasive diagnostic modality able to detect eosinophilic myocarditis at any stage. On contrary, endomyocardial biopsy is associated with risk of complications and possibility of false negative results.

Introduction

Hypereosinophilic syndrome (HES) is a rare hematologic disorder characterized by persistent overproduction of eosinophils causing multiple organ damage (1). Cardiac involvement in HES is frequent, found in approximately 50% of patients with HES, and is associated with high morbidity and mortality rates (2-4). It is caused by eosinophil infiltration of the endomyocardium with subsequent tissue damage and endomyocardial fibrosis (5). Clinical manifestations include signs of heart failure, arrhythmias, or embolic events caused by intracardiac thrombi (4, 6). We present a case of a patient with eosinophilic myocarditis clinically manifesting with progressive cardiac cachexia.

Case report

A 66-year old female patient with no previous cardiac history was admitted in a local hospital in December 2021 due to significant weight loss and symptoms of heart failure (exertional dyspnea, fatigue, lower extremity edema). Patient's initial weight of 79 kg was gradually reduced to 49 kg during past three years, with particular acceleration of weight loss in the past three months before admission. Her past medical history included bronchial asthma, chronic pansinusitis and hypothyroidism. Due to significant cachexia, patient underwent comprehensive cancer screening - with all negative results including PET/CT scan. At the same time, echocardiographic examination revealed a large right ventricular formation filling up to two thirds of the right ventricle, resembling a tumorous mass. Patient was thus referred to our medical center in order to complete the diagnostic process of presumed malignant cardiac tumor as an underlying cause of cachexia.

At admission to general cardiology department in our medical center in January 2022, asthenic patient with BMI 18 presented with holosystolic apical murmur and right-sided pleural effusion. Lower extremity edemas had already been removed after diuretic treatment. ECG at admission showed signs of subendocardial injury in anterolateral

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Received: July 20, 2022 **Published:** August 03, 2022

Citation: Elena Teringová, MD[1], Peter Penz, MD[1], Jana Poláková Mištinová, MD[2], Pavel Chňupa, MD[3], Marek Orban, MD[1]*. Eosinophilic Myocarditis presenting with progressive Cardiac Cachexia. Int Clin Img and Med Rew. 2022; 2(4): 1089.

DOI: *10.55920/IJCIMR.2022.02.001089*

leads (Figure 1).

The initial laboratory findings were as follows (Table 1): the white blood cell count was within normal limits (5.02 x 10*9/l - normal range 4.0-10.0 x 10*9/l), with boundary elevation of peripheral eosinophilia (absolute eosinophil count 0,67 x 10*9/l, normal range 0.0-0.5 x 10*9/l, relative eosinophil count 13.3%, normal range 0.0-5.0%), C-reactive protein level was increased to 51.3mg/l (normal range 1.0-5.0), level of NTproBNP was elevated to 4400ng/l (normal range 0.0-125.0), troponin level was mildly increased (high sensitive troponin T 125ng/l, normal range 0.0-14.0), D-dimer was elevated to 4.03mg/l (normal range 0.0-0.55).

Echocardiography confirmed dilatated right ventricle with a large tumorous mass, dilatated right atrium, normal left chamber size with preserved systolic function, and moderate mitral regurgitation due to retraction of posterior leaflet (Figure 2).

Targeted biopsy of the intracardiac mass was performed. The procedure was carried out in analgosedation with transesophageal echocardiography guidance. Eight samples were obtained from the apical third of the right ventricle. However, the procedure was complicated

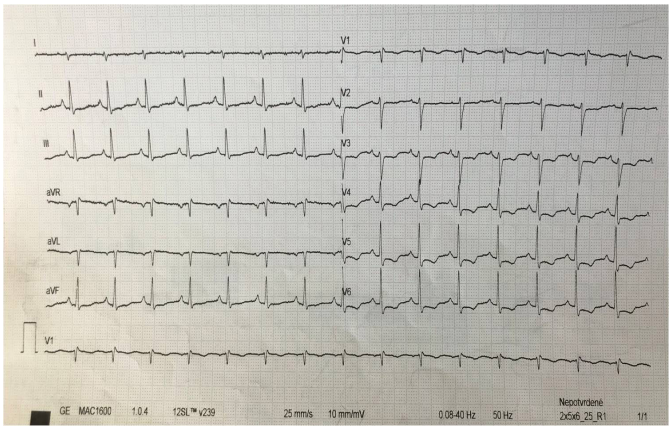

Figure 1: *Electrocardiogram showing minor ST segment depression in the anterolateral leads associated with T wave inversion*

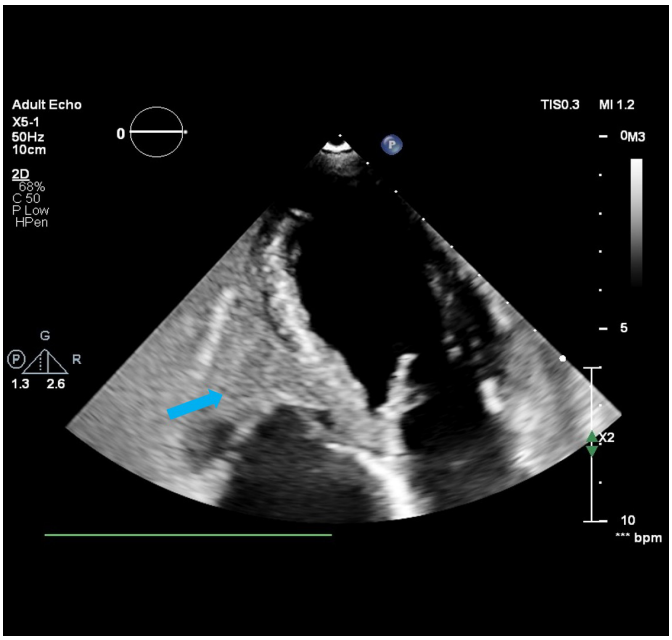

Figure 2: *Echocardiographic examination at admission showing dilatated right ventricle with a large tumorous mass in the apex filling up to two thirds of the right ventricle, dilatated right atrium, normal left chamber and left atrium size.*

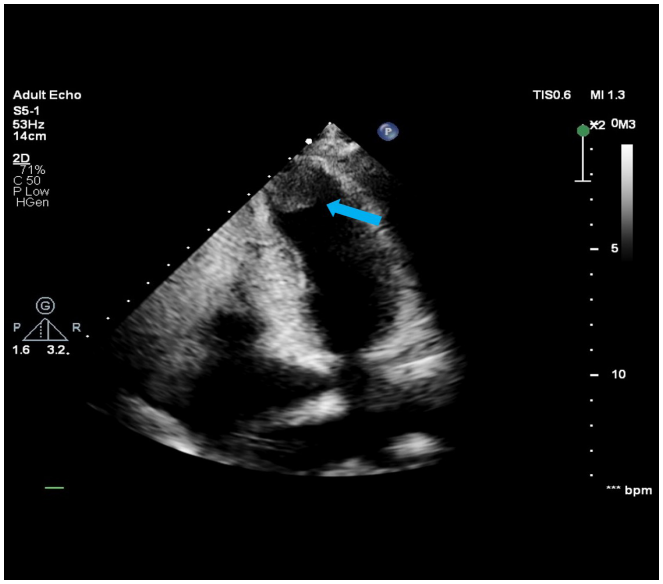

Figure 3: *Echocardiographic examination after the biopsy revealing an echodense mass also in the left ventricular apex.*

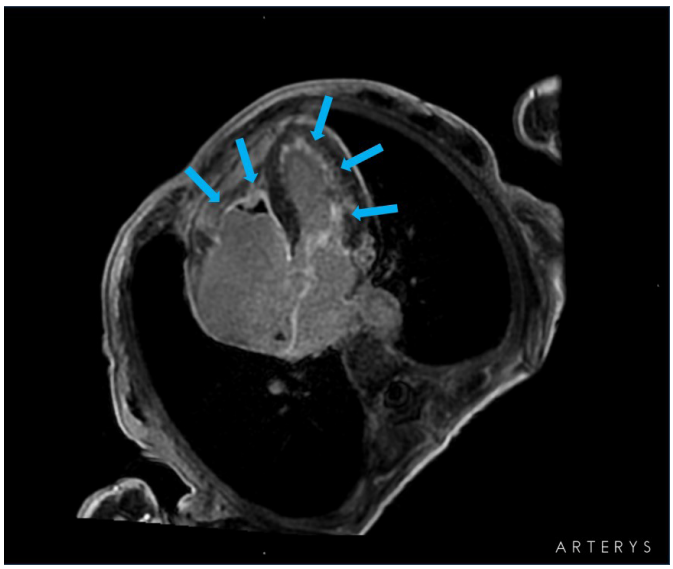

Figure 4: *Cardiac magnetic resonance imaging showing late gadolinium enhancement in apical and midventricular segments of both ventricles and in both papillary muscles.*

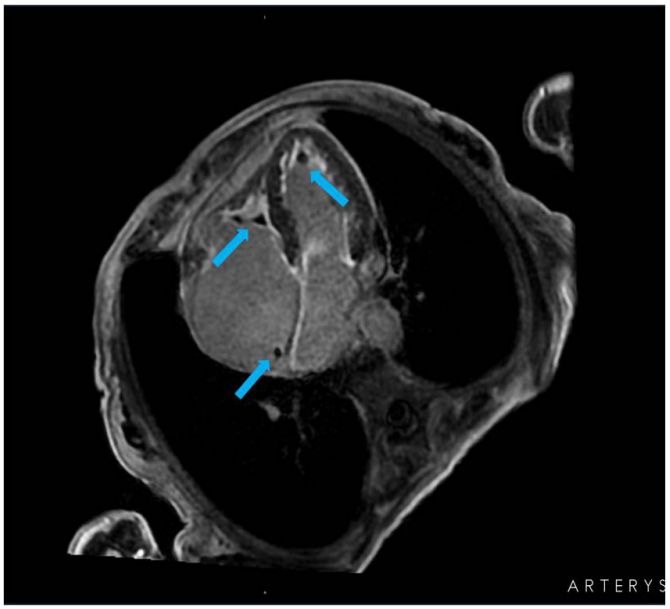

Figure 5: *Cardiac magnetic resonance imaging revealing presence of four intracardiac thrombi located in the right and left ventricular apex, right atrium and right atrial auricle.*

by right ventricular perforation causing hemopericardium and cardiac tamponade. Immediate pericardiocentesis was conducted in the cath-lab and hemopericardium was successfully removed.

Subsequently, patient was admitted to department of acute cardiology. On follow-up transthoracic echocardiography besides large right ventricle mass an echodense mass was observed in the left ventricular apex (size 12x13mm) (Figure 3). Presence of formations in both ventricular apexes, together with laboratory findings of eosinophilia and pathological EKG, led us to presumption of non-malignant disease etiology, specifically eosinophilic myocarditis.

CMR was added in order to confirm the diagnosis. Results showed late gadolinium enhancement with diffuse subendocardial distribution in apical and midventricular segments of both ventricles and in both papillary muscles (Figure 4). Further, presence of several intracardiac thrombi was detected: besides large thrombus obliterating apex and midventricular part of the right ventricle, smaller thrombi were described in left ventricular apex (9x13mm), right atrium (5x5mm) and right atrial auricle (8x6mm) (Figure 5). CMR thus confirmed distinctive signs of eosinophilic myocarditis.

Histological results from endomyocardial biopsy definitely ruled out initially presumed diagnosis of cardiac tumor, since no signs of malignant proliferation were detected. Histological evaluation of evacuated hemopericardium revealed high concentration of eosinophils (up to 50 eosinophils per one high-power field) without cytogenetic or molecular abnormalities, what represents a characteristic finding in idiopathic hypereosinophilic syndrome.

Consequently, anticoagulant and corticosteroid treatment was initiated, alongside with nutrition support and heart failure treatment. Patient was transferred back to referring hospital for further hematologic examination, including bone marrow biopsy. However, despite the treatment, patient died due to progressive irreversible cachexia with the weight of 42kg in March 2022.

Discussion

Eosinophilic myocarditis is associated with high morbidity and mortality rates. Particularly presence of intracardiac thrombi and signs of heart failure is associated with significantly poor prognosis, reaching nearly 30% mortality (6). As demonstrated also in our case, advanced stage of eosinophilic myocarditis led to progressive cardiac cachexia resulting in patient's death, despite initiated intensive treatment.

Eosinophilic myocarditis is typically associated with persistent significant hypereosinophilia with absolute eosinophil count of >1.5/nL during period exceeding 6 months (1). Level of hypereosinophilia, however, shows no correlation with the extend of endomyocardial damage. Eosinophilic myocarditis can even occur in the absence of peripheral eosinophilia (7). In agreement, our findings demonstrated only borderline peripheral eosinophilia, while CMR provided signs of an advanced stage of eosinophilic myocarditis.

Echocardiography represents first-line imaging modality for detecting eosinophilic myocarditis, however, frequently is not able to reveal early stages. In later phases, echocardiography can detect endomyocardial fibrosis, intracardiac thrombi typically located in ventricular apexes, signs of restrictive ventricular filling and/or valvulopathy. In our patient, large cardiac mass in the right ventricle was initially mistaken with malignant cardiac tumor. Presumption of cardiac tumor was strongly supported by clinical manifestation of progressive cachexia. On the other hand, laboratory findings with elevated eosinophil count, ECG and presence of formations in both ventricular apexes led us to diagnosis of eosinophilic myocarditis.

The gold standard diagnostic modality of eosinophilic myocarditis remains histological verification through endomyocardial biopsy (EMB). However, EMB represents an invasive procedure with risk of complications. As documented in our case, EMB was complicated with right ventricular perforation causing hemopericardium and cardiac tamponade. Moreover, EMB may bring inconclusive findings due to patchy pattern of tissue damage in eosinophilic myocarditis.

Finally, CMR confirmed signs of eosinophilic myocarditis with LGE found subendocardially in apical and midventricular segments of both ventricles and in both papillary muscles, along with detection of several intracardiac thrombi. Our case highlights the importance of CMR imaging in diagnostic process of eosinophilic myocarditis, since it represents a noninvasive diagnostic tool enabling detection of eosinophilic myocarditis at any stage.

Conclusion

We present a case of advanced stage of eosinophilic myocarditis clinically manifesting with progressive cardiac cachexia, terminating in patient's death despite initiated treatment. In clinical practice, presence of intracardiac formations in both ventricular apexes should raise suspicion of eosinophilic myocarditis, even in absence of significant peripheral hypereosinophilia. Our case also highlights the importance of CMR imaging in diagnostic process of eosinophilic myocarditis, since it represents a noninvasive modality able to detect even early stages of eosinophilic myocarditis. On contrary, endomyocardial biopsy is associated with risk of complications and possibility of false negative results.

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