EOSINOPHILIC MYOCARDITIS: RESPONSE TO CORTICOSTERIODS

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ABSTRACT

Eosinophilia is seen in several of the clinical condition and it affects different system of human body. Eosinophils can infiltrate in heart and causes range of cardiac abnormalities. Heart involvement usually occurs when eosinophil count exceeds 1.5×10^{9}/L for at least 6 months. Eosinophilic myocarditis is a rare condition. Few cases have been reported and most of them are from western community. The disease is potentially fatal and mortality is high if not recognized on time. Studies have shown various responses to treatment with corticosteroid and other standard heart failure measures.

We present a 35 year male that came with complains of shortness of breath and fatigability. He was diagnosed as esoniphilic myocarditis and treated with corticosteroid and other heart failure measures. The patient showed excellent response to therapy. He became entirely asymptomatic and his cardiac function (ejection fraction) became normal during follow up at 60 days.

KEYWORDS: Eosinophilia; Corticosteriods; Myocarditis

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INTRODUCTION

Eosinophilic myocarditis (EM) is a rare condition and is one of the fatal complications of hypereosonophilia. It can result from various conditions like parasite infection, allergic disease, granulomatous disease, connective tissue disease, vasculitis such as Churg-Strauss syndrome, leukemia or primary hypereosinophilic syndrome.[1,2] Clinical features are variable, ranging from mild chest pain, palpitations associated with transient ECG changes. It may be associated with shortness of breath. On the other-hand it can present as life-threatening situations like cardiogenic shock and ventricular arrhythmia.[3,4]

Due to high eosinophil in circulation, there occurs diffuse or focal eosinophilic infiltration of the myocardium. Release of eosinophilic granule proteins such as eosinophil cationic protein (ECP) and major basic protein (MBP), causes dysfunction of myocyte mitochondria leading to myocardial lesions, as well as endocardial necrosis. Acute inflammatory response with abundant eosinophils, myocytolysis may develop, as well as endomyocardial thrombi, necrotizing vasculitis, pericarditis, pericardial fibrosis and endomyocardial fibrosis can occur.[3,5]
We present a rare case of eosinophilic myocarditis who was treated with standard heart failure measures and corticosteroid who responded well with medication and achieved complete recovery of cardiac function in follow up.

CASE REPORT

A 35 years male, a driver cum farmer by profession from Rupendhai Nepal presented with complaints of shortness of breath for several months duration which was increased over last 2 weeks. It was associated with nonspecific chest pain and cough which was almost dry. He also complained of generalized weakness and fatigability for almost similar duration. He denied history of diabetes, hypertension, pulmonary tuberculosis or any other significant illness in the past. He was a non smoker and denied history of alcohol consumption.

On examination, he was dyspnic with respiratory rate of 24/min. His blood pressure was 110/60 mmHg, pulse was 110/minute and SPO_2 was 96%. He was afebrile. His respiratory, cardiovascular and abdominal examination revealed no significant abnormality.

The patient underwent routine investigations. X-ray chest was unremarkable, ECG showed sinus tachycardia with heart rate of 117 beats per minute. There was nonspecific ST-T changes which is shown in figure 1.

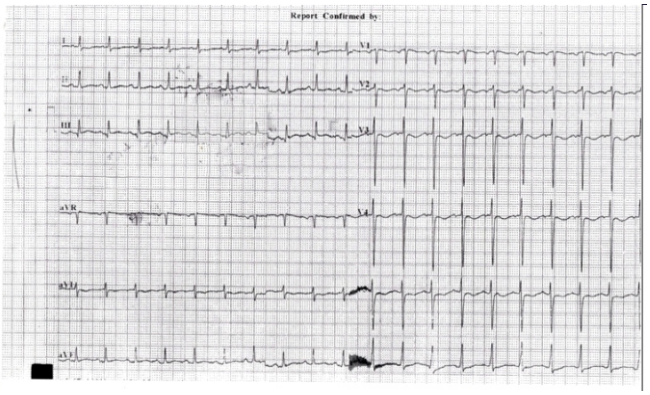

Figure 1: ECG during presentation

Complete blood count showed Hemoglobin of 18.2 gm/dl. Total count was 16,100 /mm^3 with Neutrophil 40%, Lymphocyte 10% and Eosinophil 50%. Platelet count was 2.74 lac/mm^3. His CPK MB was 93.1 IU/L and troponin I was positive. Patient underwent transthoracic echocardiographic examination which revealed dilated left atrium (LA=48 mm) and left ventricle (LV=60 mm). Both interventricular septum (IVS) and (LVPW) measured 12 mm in diastole. There was global LV hypokinesia with LV systolic dysfunction and LVEF of 45%. There was mild-moderate MR and minimal pericardial effusion. On the basis of characteristic features of heart failure, presence of eosinophilia in blood and echocardiographic findings, eosinophilic myocardit was diagnosed.

The patient was hospitalized. He underwent treatment with injection methylprednisolone 1 gm IV for 3 days followed by oral prednisolone 1mg/kg. Diethyl carbamazine (DEC) was started. Other medication started were telmisartan 40 mg/OD, inj furosemide followed by furosemide+spironolactone tablet, bisoprolol and other supportive measures. During mean time patient underwent serial ECG monitoring, which revealed no significant changes. Peripheral smear showed no atypical cells. His P- ANCA, C- ANCA and microfilaria tests were negative. CT scan of the chest revealed minimal pleural and pericardial effusion.

The patient started improving from the 2nd day of medication and recovered almost completely clinically and discharged after 7 days of hospitalization. His repeat CBC revealed normal total WBC count with 8% eosinophils. The patient underwent coronary angiogram, which revealed normal coronaries.

The patient was treated with Beta blocker, ARB, diuretics and predinosolone. Prednisolone was gradually tapered and kept in 5mg/ day. At 60 days follow up he was completely asymptomatic and could join his daily work comfortably. His

echocardiogram in follow up was almost normal. The comparison of echocardiogram finding during presentation and at follow up is shown in table 1.

Table 1: Comparison of echocardiogram between 1st presentation and at 60 days follow up

Parameters	During 1st presentation	At follow up
Chamber Size	Dilated LA/LV	Normal
Wall Motion	Global LV hypokinesia	Normal
LVEF	45%	60%
Thickness	IVS/ LVPW= 12 mm	IVS/ LVPW= 10 mm
MR	Mild- moderate	Trace

DISCUSSION

Wilhelm Löffler described originally the cardiac infiltration of eosinophil in 1936.[6,7] Eosinophilic myocarditis is a rare variant of myocarditis characterized by focal or diffuse myocardial inflammation with infiltrating eosinophils. To date there are less than 30 published case reports of EM. Due to its rarity and high fatality rate, it is often under-recognized and may be first evident on postmortem examination.[8,9,10] Five year mortality of the disease is around 30%.[3] The disease may be evident at any age. This subject was 35 years old. Studies have shown that disease may present from age group of 2-83 years.[8]

The gold slandered diagnostic test for EM is endomyocardial biopsy (EMB). But in many circumstances the procedure is not readily available or the patient may not be ready for the consent and diagnoses of eosinophilic myocarditis may often be challenging.[11] However, under-diagnosis and late diagnosis of eosinophilic myocarditis may result in fatal outcomes such as heart failure, arrhythmias, or sudden death. The diagnostic criteria proposed by the Japanese Circulation Society Task Force may be practically useful for early recognition and treatment. Essential diagnostic features include blood eosinophilia >500 μL, cardiac symptoms, elevated cardiac enzymes, ECG changes, and abnormal echocardiography, in the setting of unremarkable coronary angiography.[12] In our case EMB was not possible. As we had strong suspicion for the EM, we initiated therapy without delay. Several studies have been reported previously where therapy was initiated in timely manner if sufficient evidence was present even if biopsy was not available.[2,9] Moreover, a biopsy is not very sensitive (50%) as the infiltrate is often focal.[13,14]

No randomized controlled trial has yet evaluated the treatment of eosinophilic myocarditis.[2] In a case report, intravenous methylprednisolone followed by 1 mg/kg/day oral prednisone, with gradual tapering showed improvement in symptoms and LVEF.[15] It was in consistence with our findings. The primary cause of eosinophilia needs to be searched and treated if possible. We also initiated and continued the Angiotensin Recepter Blocker (ARB), Beta blockers and diuretics. After 60 days of follow up, beside ARB, Beta blockers and diuretics we have kept patients on low dose of steroid with regular follow up. The duration of corticosteroids therapy in EM and the necessity of maintenance therapy needs to to be recommended through large well-designed trails. In several studies Immuno suppressive agents, such as azathioprine, mycophenolate, and intravenous gamma globulins, have been used in conjunction with steroids.[9,16]

CONCLUSION

Eosinophilic myocarditis is a rare disease with high mortality. Early diagnosis and prompt initiation of therapy is rewarding. Esonophilia in peripheral blood, raise in cardiac enzymes, changes in ECG and characteristic echcardiographic findings on the background of normal coronary angiogram is generally sufficient for the diagnosis though EMB is the gold slandard. This patient responded well with steroid and routine heart failure measures. He was completely recovered clinically and cardiac function was normal at 60 days follow up.

REFERENCES

1. *Ommen SR, Seward JB, Tajik AJ. Clinical and echocardiographic features of hypereosinophilic syndromes. Am J Cardiol 2000; 86:110-3. http://dx.doi.org/10.1016/S0002-9149(00)00841-9*

2. *Liao YC, Su CS, Teng CL, et al. Acute necrotizing eosinophilic myocarditis in a young woman. Journal of the Chinese Medical Association 75 (2012) 536-538. http://dx.doi.org/10.1016/j.jcma.2012.07.011 PMid:23089407*

3. *Li H, Dai Z, Wang B, et al. A case report of eosinophilic myocarditis and a review of the relevant literature. BMC Cardiovascular Disorders. 2015; 15:15. http://dx.doi.org/10.1186/s12872-015-0003-7, PMid:25887327 PMCid:PMC4359588*

4. *Caforio AL, Pankuweit S, Arbustini E, et al. Current state of knowledge on aetiology, diagnosis, management, and therapy of myocarditis: a position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J. 2013; 34(33)):263648.*

5. *Hara T, Yamaguchi K, Iwase T, et al. Eosinophilic myocarditis due to Churg-Strauss syndrome with markedly elevated eosinophil cationic protein. Int Heart J. 2013; 54(1):513. http://dx.doi.org/10.1536/ihj.54.51 PMid:23428926*

6. *Löffler W. Endocarditis parietalis fibroplastica mit Bluteosinophilie, ein eigenartiges Krankheitsbild. Schweiz Med Wochenschr. 1936; 66: 817820.*

7. *Reuss CS, Wilansky S. Eosinophilic Heart Disease in Acute Myeloproliferative Disorder. Circulation. 2007; 115: e614-e616. http://dx.doi.org/10.1161/CIRCULATIONAHA.107.692574 PMid:17562961*

8. *Kawano S, Kato J, Kawano N, Yoshimura Y, Masuyama H, Fukunaga T, Sato Y, Maruyama H, Mihara K, Ueda A, Toyoda K, Imamura T, Kitamura K: Clinical features and outcomes of eosinophilic myocarditis patients treated with prednisolone at a single institution over a 27-year period. Intern Med 2011; 50:975981. http://dx.doi.org/10.2169/internalmedicine.50.4079*

9. *Rizkallah J, Desautels A, Malik A, et al. Eosinophilic myocarditis: two case reports and review of the literature. BMC Research Notes 2013; 6:538. http://dx.doi.org/10.1186/1756-0500-6-538, PMid:24344829 PMCid:PMC3878496*

10. *Amini R, Nielsen C. Eosinophilic myocarditis mimicking acute coronary syndrome secondary to idiopathic hypereosinophilic syndrome: a case report. J Med Case Reports 2010; 4:40. http://dx.doi.org/10.1186/1752-1947-4-40, PMid:20181108 PMCid:PMC2830978*

11. *Sohn KH, Song WJ, Kim BK, et al. Eosinophilic myocarditis: case series and literature review. Asia Pac Allergy 2015; 5:123-127. http://dx.doi.org/10.5415/apallergy.2015.5.2.123 PMid:25938077 PMCid:PMC4415178*

12. *Japanese Circulation Society Task Force Committee on Acute and Chronic Myocarditis. Guidelines for diagnosis and treatment of myocarditis (JCS 2009) [Internet]. Tokyo; Japanese Circulation Society; [cited 2015 August 5]. Available from: http://www.j-circ.or.jp/ guideline/pdf/JCS2009.*

13. *Burke AP, Saenger J, Mullick F, et al. Hypersensitivity myocarditis. Arch Pathol Lab Med 1991; 115(8):764-769. PMid:1863186*

14. *Amini R, Nielsen C. Eosinophilic myocarditis mimicking acute coronary syndrome secondary to idiopathic hypereosinophilic syndrome: a case report. Journal of Medical Case Reports 2010; 4:40. http://dx.doi.org/10.1186/1752-1947-4-40; PMid:20181108 PMCid:PMC2830978*

15. *Corradi D, Vaglio A, Maestri R, et al. Eosinophilic myocarditis in a patient with idiopathic hypereosinophilic syndrome: Insights into mechanisms of myocardial cell death. Hum Pathol 2004; 35:1160-3. http://dx.doi.org/10.1016/j.humpath.2004.05.008; PMid:15343520*

16. *Ali AM, Straatman LP, Allard MF, et al. Eosinophilic myocarditis: Case series and review of literature. Can J Cardiol 2006; 22(14):1233-1237. http://dx.doi.org/10.1016/S0828-282X(06)70965-5*