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HYPEREOSINOPHILIA PRESENTING AS RESTRICTIVE CARDIOMYOPATHY - LOEFFLER'S ENDOCARDITIS:-

General Medicine

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ABSTRACT

Loeffler's endocarditis is a rare restrictive cardiomyopathy caused by abnormal endomyocardial infiltration of eosinophils, with subsequent tissue damage from degranulation, eventually leading to fibrosis. We present a unique case of a woman who presented with signs and symptoms of heart failure was found to have hypereosinophilia and restrictive cardiomyopathy diagnosed with Loeffler's endocarditis. high-dose corticosteroids were used to reverse the cardiac injury and to improve the clinical outcome.

KEYWORDS

INTRODUCTION

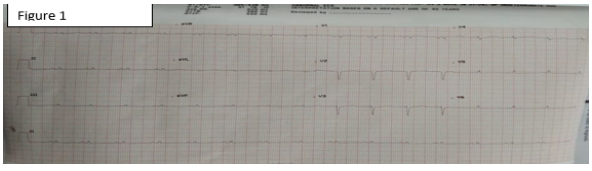
Hypereosinophilia is defined by the presence of ≥ 1500/mm^3 eosinophils in the peripheral blood, and may be reactive, neoplastic, or idiopathic.[1,2] A marked and persistent overproduction of eosinophils that subsequently infiltrate and damage multiple organs via a toxic protein is referred to as hyper eosinophilic syndrome (HES). The age-adjusted incidence rate of HES is approximately 0.036 per 100,000 person-years.[3] The clinical manifestations of HES depend on involment of either the heart, lung, nervous system or bone marrow which targeted by the eosinophils. Cardiac manifestations occur due to acute eosinophilic injury in the endocardium in half of patients with HES and are a major cause of morbidity and mortality. Cardiac inflammation is reveled by evidence of endomyocardial fibrosis is known as "Loeffler's endocarditis."[4]

We report a case of a 45-year-old female diagnosed with left-sided restrictive cardiomyopathy with a preserved ejection fraction, which was identified as Loeffler's endocarditis.

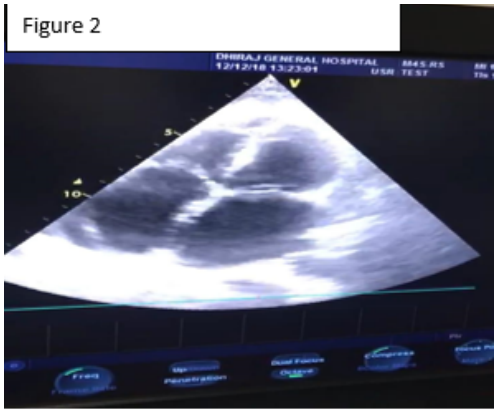
CASE

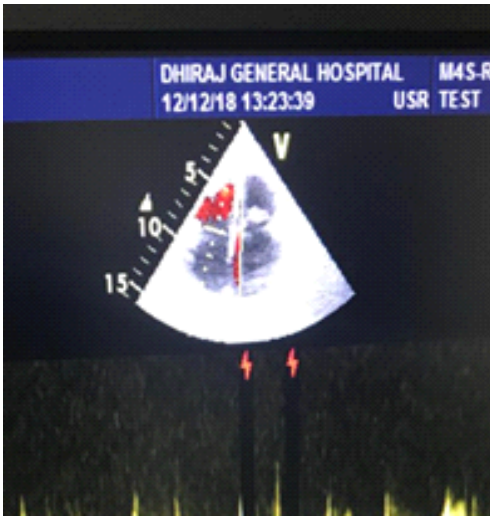
A 45 -year-old female with no co-morbidities presented to our institution with progressively worsening dyspnea on exertion and paroxysmal nocturnal dyspnea for duration of 30 days, She also had facial puffiness with abdominal distension and pedal edema associated with low grade fever since 10 days The physical examination showed pallor, raised JVP (> 5cm above sternal angle). Cardiovascular system examination was suggestive of pansystolic murmur in mitral area. Respiratory examination was suggestive of bilateral crackles at the lung bases. Per abdomen examination revealed hepatosplenomegaly .
The complete blood count at presentation showed Hemoglobin 7.9 gm% and white blood cell count 59300 cell/cumm with 13% neutrophils, 6% lymphocytes, and 80% eosinophils with absolute eosinophil counts 47400/mL (normal 20–520/ml). Peripheral smear of blood suggested microcytic hypochromic RBCs with target cells, tear drop cells and elliptocytes , leukocytosis with marked eosinophilia, eosinophils showing multilobation, hypolobation and vacuolation and platelets were adequate.
ECG showed normal sinus rhythm and non specific ST-T changes.

Echocardiography revealed that both atriums were enlarged and a restrictive left ventricle (LV) filling pattern with a large LV clot in midcavity was noted . Both ventricles displayed normal dimensions in the absence of regional wall motion abnormalities, and both ventricular systolic functions were preserved (left ventricular ejection fraction 55%). pericardial effusion was observed on lateral surface and right ventricle side with no signs of tamponade. An additional, extensive workup of secondary causes of eosinophilia proved negative and excluded other causes, leading to the final diagnosis of Loeffler's Endocarditis with Restricted Cardiomyopathy.



In above figure 1 ECG showed normal sinus rhythm and non specific ST-T changes.





In above figure 2 Echocardiography showing restrictive cardio myopathy. An apical four chamber view is showing biventricular enlargement with normal LV thickness and LV clot

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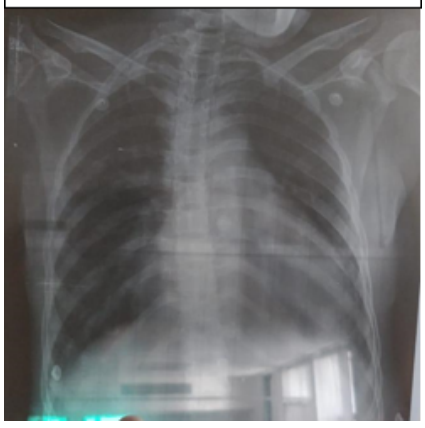
Figure 3



In above Figure 3 X-ray chest show cardiomegaly and did not show any pulmonary infiltrate as absolute eosinophil count was very high.

Treatment was started immediately with Nicoumalone 2mg once daily and a high dose of prednisone (1 mg/kg/d) which was gradually tapered and discontinued after 4 weeks. Over the next few days, patient began to feel much better, demonstrating marked improvement in her symptoms, with no complaints of shortness of breath on exertion. Eosinophil counts also normalized. Her congestive cardiac failure subsided and hepatosplenomegaly diminished in size with improvement in haemoglobin level.

DISCUSSION

Loeffler's endocarditis is rare type of restrictive cardiomyopathy, presenting late when patient has already developed heart failure.[5] In early phase of the disease, presentation is subtle, with nonspecific features like fever, malaise, chest pain and discomfort. During this period the eosinophilic infilteration of myocytes starts, leading to release of reactive oxygen species and major basic protein from eosinophils causing injury to myocytes eventually leading to fibrosis resulting in restrictive cardiomyopathy.[6]

These patients have state of localized hypercoaguability, due to interaction between cationic proteins released from eosinophils and anionic endothelial thrombomodulin hence making these patients prone to thrombus formation in heart cavity.[7] Although it is an uncommon entity, the patients with Loeffler endocarditis can be closely monitored to prevent fibrosis because once fibrosis occurs, management becomes more difficult, usually requiring surgical intervention.[5]

Early diagnosis and high-dose corticosteroids are the cornerstone of treatment, and, may lead to restoration of cardiac function with full recovery.[6]

It is a lethal disease which eventually leads to right heart failure needing prompt diagnosis and treatment with careful monitoring.

REFERENCES

[1] Gotlib J. World Health Organization-defined eosinophilic disorders: 2015 update on diagnosis, risk stratification, and management. Am J Hematol 2015;90:1077–89. [PubMed]

[2] Gotlib J. World Health Organization-defined eosinophilic disorders: 2017 update on diagnosis, risk stratification, and management. Am J Hematol 2017;92:1243–59. [PubMed]

[3] Crane MM, Chang CM, Kobayashi MG, et al. Incidence of myeloproliferative hypereosinophilic syndrome in the United States and an estimate of all hypereosinophilic syndrome incidence. J Aller Clin Immunol 2010;126:179–81. [PMC free article] [PubMed]

[4] Tai PC, Ackerman SJ, Spry CJ, et al. Deposits of eosinophil granule proteins in cardiac tissues of patients with eosinophilic endomyocardial disease. Lancet (London, England) 1987;1:643–7. [PubMed]

5) Alam A. Various toxicities: case report. Reactions. 2017 Dec;1680:178-2.

6) Li, H., Dai, Z., Wang, B., & Huang, W. (2015). A case report of eosinophilic myocarditis and a review of the relevant literature. BMC cardiovascular disorders, 15, 15. doi:10.1186/s12872-015-0003-7.

7) Slungaard A, Vercellotti GM, Tran T, Gleich GJ, Key NS. Eosinophil cationic granule proteins impair thrombomodulin function. A potential mechanism for thromboembolism in hypereosinophilic heart disease. J Clin Invest. 1993 Apr;91(4):1721-30. PubMed PMID: 8386194; PubMed Central PMCID: PMC288152.