# Endomyocardial Fibrosis in non-tropical patient with no obvious eosinophilia presented with chest pain mimicking ACS and left ventricular thrombus

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**Abstract**

Endomyocardial fibrosis is a disease that causes restrictive cardiomyopathy, it varies in presentation ranging from asymptomatic to full-blown heart failure. its pathogenesis is poorly understood and it poses a diagnostic and treatment challenge due to its infrequent occurrence.

**Endomyocardial Fibrosis in non-tropical patient with no obvious eosinophilia presented with chest pain mimicking ACS and left ventricular thrombus**

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**Key clinical message:**

Endomyocardial fibrosis is a rare cause of restrictive cardiomyopathy, its pathology is poorly understood, and further theories are proposed as a cause. Diagnosis and its treatment are challenging, and further studies are needed to expand on it.

**Abstract**

Endomyocardial fibrosis (EMF) is a disease that known to cause restrictive cardiomyopathy. It shows high prevalence in tropical country. Several triggering factors has been proposed, however, the pathogenesis is still mystery. The disease is progressive in nature and the outcome is generally unfavorable. Most common symptoms is heart failure, however, atypical presentation may expected.

Our case presented with symptoms suggestive of ischemia and missed diagnosed initially as ischemic cardiomyopathy. The aim of this report is to increase the attention and awareness for this disease.

We present a case of a 53-year-old man referred to emergency department for sudden chest pain, left sided and non-radiating lasted for several minutes, awoke him from sleep with no associated symptoms. He is known to have Diabetes type-2 and hypertension on oral therapy. Cardiac markers were within normal limit. The patient discharged home with appointment to cardiology out-patient clinic. Echocardiography done and revealed mildly reduced left ventricular (LV) systolic function Ejection Fraction of 46 %, asymmetric LV hypertrophy affecting the apical segments with aneurysm and calcified apical thrombus. CT coronary angiography done with non-significant Left Anterior Descending artery lesions and left ventricular hypertrophy affecting the apex with calcified apical thrombus. Further investigation by cardiac MRI revealed apical thrombus and late apical uptake suggesting Endomyocardial Fibrosis of possible eosinophilic etiology. The patient continued to have attacks of similar chest pain for which stress cardiac MRI done and was negative for ischemia. Other diagnostic work-up was done including hematological and serological tests such as Antinuclear Antibodies and Schistosoma Antibodies.

The patient was put on valsartan and Bisoprolol with oral anticoagulant (vitamin K antagonist) and Rosuvastatin.

Conclusion: EMF may have heterogeneous presentation and should be considered in patient with calcific apical thrombus without previous history of cardiac problem, even in non-tropical region

**Introduction:**Endomyocardial fibrosis (EMF) is a progressive restrictive cardiomyopathy that usually affects the left, right, or both ventricles. It can also affect the outflow of the ventricles, leading to the development of symptoms of failure of the involved ventricle (1). The restrictive involvement is usually seen because of apical fibrosis which usually arises from collagen deposition and fibroblast proliferation. It is prevalent in tropical and subtropical parts of the world. The main cause of it is still unknown, however, there are multiple theories available regarding its development (2). Treatment is challenging as currently there is limited data, medical therapy as a case of restrictive heart failure might offer a symptomatic relief, however the disease is usually progressive, and the long-term outcome is generally poor. On the other hand, surgical resection of the endocardium might offer a definite treatment however there is still a risk of recurrence, and it carries a high mortality rate (3).

We report a patient who presented with chest pain and on subsequent investigations were diagnosed with EMF. The patient has frequent visit to Heart failure clinic, and he is doing well.

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**Case presentation**

A 53-years-old male patient from the Eastern Mediterranean Region with background of diabetes mellitus type II (DM II) and hypertension (HTN), both controlled with oral medications, presented to the primary health care center because of acute onset chest pain. Chest pain lasted for several minutes and was left-sided and non-radiating. There were no associated symptoms and no specific worsening or relieving factors. Family history is non-significant. On examination patient was afebrile, normal pulse and respiratory rates (HR 78, RR 16), blood pressure was high at 155/94 mmHg. Systemic examination was unremarkable. ECG (**Figure 1** ) showed sinus rhythm with T-inversion in I, II, avl, V3-V6, which were considered old when compared to the ECG in 2017. The Patient was transferred rapidly to the emergency department (ED) for further evaluation and management through the emergency ambulance service.

In the ED, the patient was kept under observation, he was chest pain free with stable vital signs and physical examination. Chest radiographs and laboratory investigations were unremarkable including for two sets of high sensitivity cardiac troponin-T (Troponin T HS) with values of 7 ng/L and 8 ng/L respectively. Rest of the laboratory panel are as mentioned in **table 1** . The patient was discharged with cardiology outpatient follow up advise.

A 2D transthoracic Echocardiography was done four days later during the outpatient cardiology clinic follow up visit, (**Figure 2** ), which revealed; mildly reduced left ventricular (LV) systolic function with Ejection Fraction of 46 %, asymmetric LV hypertrophy affecting the apical segments which was aneurysmal. The apical cap contained a calcific material (calcified aneurysm vs calcified thrombus) and Grade 1 diastolic dysfunction were noted. Further evaluation with CT coronary angiogram showed non-significant left anterior descending artery lesion with concentric LV myocardial hypertrophy affecting the LV apex with subendocardial apical calcifications.

Cardiac MRI (CMR) images were performed later. Images obtained on a 1.5 tesla scanner (Philips Ingenia) revealed thickening of the akinetic LV apical segments (**Figure 3** ) which demonstrated non-ischemic intense LV apical sub-endocardial late gadolinium enhancement (**Figure 4** ). An apical LV thrombus was also noted. CMR concluded that the findings were consistent with EMF.

At this point, a diagnosis of EMF was made, and the patient was started on therapeutic dosing of anticoagulation in addition to Valsartan and Bisoprolol. Blood works were repeated, C-reactive protein level was 4.9 mg/L and ANCA and Anti-Schistosoma-Ab were negative. Peripheral smear revealed normochromic, normocytic RBC's with few teardrop cells and mild rouleaux formation seen with a mild absolute eosinophilia and occasional reactive lymphocytes. The patient continued to have a non-exertional, non-radiating, on and off left sided chest pain, hence an adenosine stress CMR perfusion study was performed. This CMR performed almost 6 weeks after the initial CMR showed no stress induced perfusion defects, but there was regression of the size of LV apical thrombus with no other obvious interval significant changes.

**Discussion:** EMF is a rare and a newly emerging entity, it was first described in 1947 in Uganda. It is more commonly found in the developing world, such as Africa, Asia, and South America (1). it is characterized mainly by fibrosis of the apical endocardium mainly in the right, left or both ventricles. The clinical manifestations of it usually arise from the symptoms of heart failure secondary to restrictive LV filling (2). The symptoms depend on the extent of involvement of the ventricles. The initial phase of the disease is acute carditis, characterized by febrile illness and in some cases, it progresses to cardiogenic shock. But the vast majority present during the chronic phase with symptoms of heart failure, arrhythmia and/or thromboembolism (1, 4).

Generally, the main pathological event is unknown, however there are multiple theories about its development and further studies are needed to further unfold its etiologies and help us understand it in a better way. some of the theories that have been postulated are: 1- eosinophilia related process, which can lead to endocardial damage and fibrosis. 2- infection with prevalent organisms however no specific organism has been linked to its development. 3- autoimmune phenomenon, has also been considered as some studies have found anti-myosin antibodies, however finding these antibodies is not specific, and lastly 4- genetic components have been suggested as well, but none were completely linked to it (5). Diagnosis is usually challenging, as little is known about the true nature of the disease, however there are several supporting features that we can depend on, ECG is normal in most cases, however, in advanced cases it can show non-specific ST-T wave abnormalities, variable degrees of conduction block, or arrythmias (6). Complete blood count can show eosinophilia which can further support the diagnosis; but it is usually absent (7). Echocardiography is the modality of choice while evaluating for the disease, it can show apical obliteration, endocardial surface thrombi, or AV valve abnormalities (8). Other imaging modality that is currently gaining popularity is Cardiac MRI with gadolinium contrast enhancement which can demonstrate enhancement that further supports the diagnosis (9).

Treatment is often challenging, as there is little evidence on how to treat. Standard medical therapy is to be

used, such diuretics for decongestion, ACEi/ARBs and beta blockers which can provide symptomatic relief, but the disease usually progresses in a short period of time. Anticoagulation is indicated for patients with proven endocardial thrombus (10). Surgery is one of the modalities that has improved survival in patients with advanced disease, the currently used approach includes endocardiectomy combined with valvular replacement if the valve is damaged, however it conveys a high mortality rate approaching 20%, in addition, recurrence after surgery has also been described. In severe advanced cases resistant to treatment, last resort is heart transplantation (11). Prognosis is usually poor due to the progressive nature of the disease and the increased risk for complications such as, arrythmias and sudden cardiac deaths (12).

**Data availability statement:**The data that support the findings of this study are available from the corresponding author upon reasonable request.

**Conflict of interest:** The authors have no conflict of interest to declare.

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**Ethical Approval**

The case report was approved by Hamad medical corporation, MRC number MRC-04-21-242.

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