https://doi.org/10.3329/cardio.v16i2.75251

Case Report

Eosinophilic Granulomatous Polyangitiis- a Hopeful Outcome of a Sinister disease- A Case Report

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Key Words : *Myocarditis, Eosinophilic Granulomatous Polyangitiis.*

Abstract:

Eosinophilic myocarditis is a rare form of myocardial inflammation characterized by eosinophilic infiltration and often accompanied by eosinophilia. It can be fatal in in its initial presentation. Although the condition is rare, it's quite variable in terms of clinical presentation and prognosis ranging from fulminant myocarditis to uneventful recovery.

We present a case of a young female patient with eosinophilic myocarditis who had favorable outcome after treatment with steroid and immunosuppressive agents.

(Cardiovasc j 2024; 16(2): 102-104)

Case Report:

37 years old female with history of asthma till age 15 treated intermittently with tapering dose of prednisone which she would need almost every year followed by upper respiratory tract infection. She was also maintained on montelukast. She also has sinonasal issues but not destructive sinonasal disease. On July 29th she developed chest pain with radiation to her left shoulder, tingling in her jaw and blurry vision which has been going on for about last 4 days before presentation. Her EKG showed diffuse ST elevation and her Troponins were elevated. She underwent cardiac catheterization which showed normal coronaries. Echocardiogram showed LVEF=44% with global hypokinesia. Cardiac MRI showed patchy enhancement with diffuse myocardial involvement suggestive of myocarditis. CXR and CT chest showed airspace opacification of upper lobes, left lower lobe and right middle lobe. CT sinuses showed left maxillary sinusitis and right mastoid opacity.

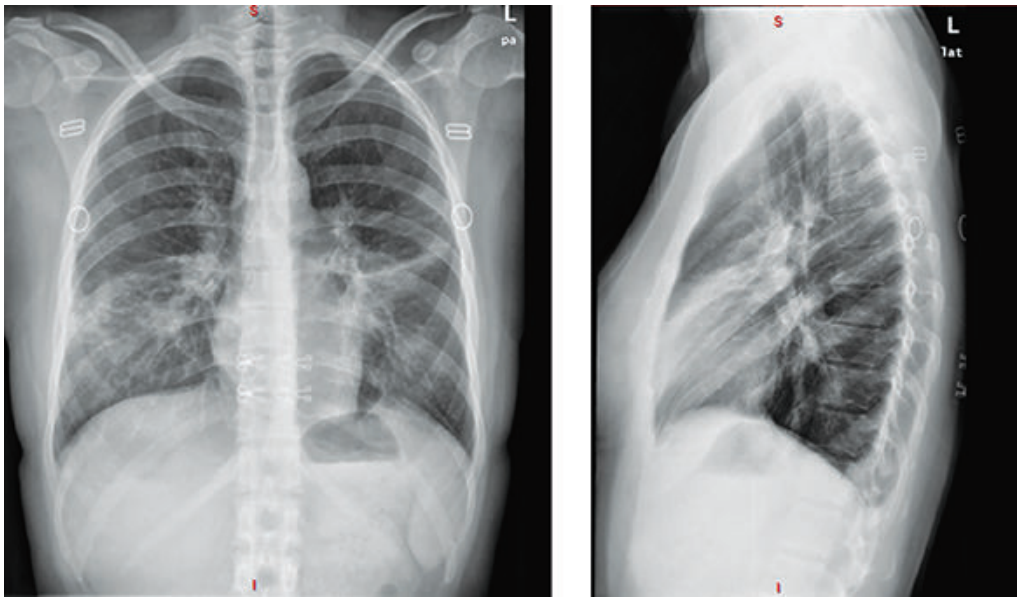

Figure 1: *Chest X -ray showing opacities in both lung fields.*

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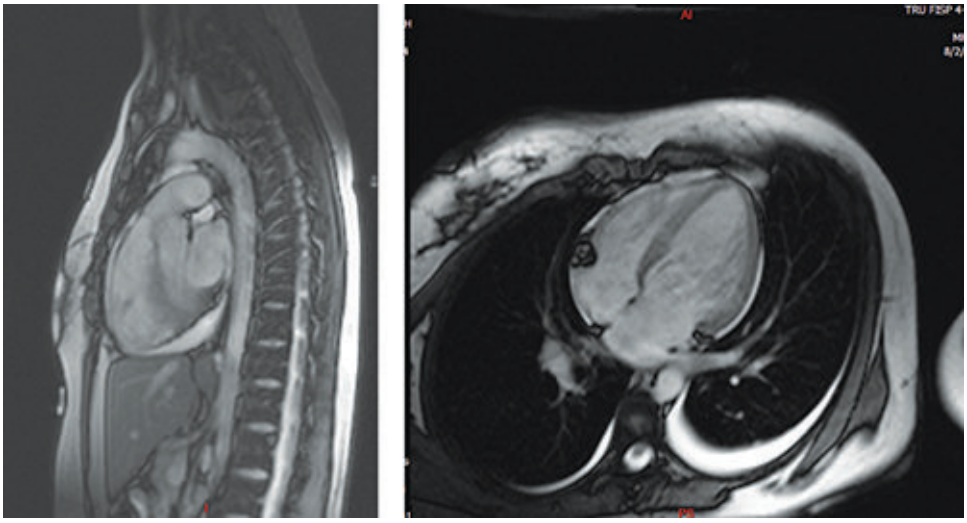

Figure 2: *Cardiac MRI showing features of myocarditis.*

Patient was treated with high dose steroid and discharged home on Prednisone 40 mg po qd. Due to her presentation with pulmonary infiltrates, significant eosinophilia and myocarditis and no infectious agent was identified she was diagnosed with Eosinophilic granulomatosis with polyangiitis (EGPA) by rheumatologist and subsequently treated with Cyclophosphamide 15/kg every 3 weeks for 3 months. Subsequently she was treated with Imuran starting with 75 mg po qd and titrated up to 200 mg/day with normalization of eosinophils and CRP and resolution of her symptoms. Her LV systolic function improved with normal LVEF up to 65%. Repeat MRI was performed which showed improvement compared to previous study. It showed LV scar only 4% and no evidence of myocardial edema.

Based on clinical and radiological improvement in her clinical status she is planning for her second pregnancy.

Discussion:

Eosinophilic myocarditis is a rare form of myocardial inflammation characterized by eosinophilic infiltration and often accompanied by eosinophilia. A systematic review of this disease entity was published for the first time in JACC 2017 titled "Eosinophilic Myocarditis: Characteristics, Treatment and Outcome".[1] In this report, main symptom on presentation was dyspnea in about 60% of patients, with peripheral eosinophilia observed in 76% of patients. Average Left ventricular ejection fraction at presentation was 35%. It was associated mainly with hypersensitivity reaction followed by eosinophilic granulomatosis and third polyangitis (formerly Churg-Strauss syndrome). These three clinical entities accounted for 50% of cases. In vast majority of cases, cause remains unknown. Clinical presentation may be variable ranging from asymptomatic to acute fulminant eosinophilic necrotizing myocarditis or chronic restrictive cardiomyopathy (Loeffler's syndrome). Often eosinophilic myocarditis can be fatal in in its initial presentation. This is usually suspected when patient presents with eosinophilia, increased Troponin and evidence of myocarditis or cardiomyopathy by MRI. Definitive diagnosis is made by myocardial biopsy.

In terms of management steroids were administered in majority of patients and temporary circulatory support was instituted in 17% of patients. Hospital mortality was 22% with highest occurrence in hypersensitivity form as a result of antibiotic or central nervous system agent.

In summary eosinophilic myocarditis has poor prognosis in acute phase. 75% patients have eosinophilia and 25% do not. Diagnosis should be made with MRI or biopsy. Dr Leslie Cooper from Mayo Clinic, Florida department of Medicine commented in his editorial regarding this entity and his title was 'importance of awareness'. Hypersensitivity myocarditis is poorest in terms of prognosis. Eosinophilic myocarditis and

polyangitis go together. Eosinophilic lesions in the liver and blood vessels called complex hypereosinophilic syndrome can be seen. Intracardiac thrombus can be seen. Use of steroid is controversial and use of antileukine antibody targeting eosinophilic production is being used. Some recover completely, some evolve onto chronic stage of left ventricular systolic dysfunction and others evolve into heart failure with preserved ejection fraction. Although the condition is rare, it's quite variable in terms of clinical presentation and prognosis. In acute fulminant forms, prognosis is poor. Multicenter trials and registry are needed to improve prognosis in such patients

Endomyocardial biopsy remains the cornerstone for diagnostic testing in eosinophilic myocarditis cases. CMR holds promise as a screening tool in suitable clinical contexts. Concerning treatment, there lacks consensus on the ideal corticosteroid dosage. Substantial clinical trials are necessary to delve deeper into CMR's diagnostic potential in EM and to determine optimal steroid regimens for its treatment.[2]

In eosinophilic myocarditis cases where feasible, CMR imaging efficiently reveals common myocarditis indicators such as myocardial edema and injury, along with ventricular dysfunction and pericardial effusion. Additionally, the magnitude of myocardial late gadolinium enhancement (LGE) corresponds to myocardial histology, where extensive LGE presence indicates necrotizing eosinophilic myocarditis. Nevertheless, CMR findings lack specificity for eosinophilic myocarditis diagnosis, necessitating myocardial biopsy and histopathology to differentiate it from other myocarditis types.[3]

Eosinophilic myocarditis is indeed a challenging condition to diagnose and manage, and cardiac MRI plays a pivotal role in its evaluation. The ability of MRI to assess various aspects such as ventricular thrombus, cavity obliteration, and fibrosis provides valuable information for both diagnosis and prognosis.

The utilization of Late Gadolinium Enhancement (LGE) sequences is particularly crucial in distinguishing between early necrotic and late fibrotic stages of the disease. This differentiation can be instrumental in guiding therapeutic decisions and assessing treatment response.

There was another case report of hopeful outcome of eosinophilic myocarditis in a 14 years old boy where it sounds like the patient had a left ventricular apical soft tissue mass, possibly indicative of eosinophilic myocarditis, given the elevated eosinophil count and the response to prednisolone treatment. The initial MRI findings showed obliteration of the apex and characteristic patterns on the late gadolinium enhancement (LGE) images. However, the patient was lost to follow-up for eight months. Upon the patient's return, a repeat cardiac MRI showed a significant reduction in the soft tissue mass and complete resolution of the inflammation, as indicated by the absence of residual LGE. This suggests a favorable response to treatment.

Eosinophilic myocarditis can be a challenging condition to manage, but this case demonstrates the potential for improvement with appropriate therapy, highlighting the importance of follow-up care.

Top of Form

In presenting our case report, we wanted to share with our physician colleagues the multiple forms of a complex disease involving multiple organs having sinister effect on patient outcomes specially when it causes fulminant myocarditis while on the other hand it may have favorable outcome with rigorous treatment protocol as was evident in our case. We also want to highlight the utility of imaging studies such as MRI in identifying initial eosinophilic myocarditis and serial study showing improvement with therapy as outlined in our case report.

Conflict of Interest - None.

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