Introduction

Eosinophilic granulomatosis with polyangiitis, previously referred to as Churg–Strauss syndrome, is a rare amalgam of systemic ANCA associated necrotizing vasculitis and a hyper eosinophilic disorder with frequent lung involvement. Most patients with EGPA present with asthma like symptoms and may be misdiagnosed with refractory asthma. Additionally, most patients with EGPA commonly die from cardiac causes even though clinically evident cardiac manifestations are uncommon.

Case Report

29-year-old male with past medical history significant for asthma, recurrent pneumonia presented with shortness of breath, sub-sternal chest pain and bilateral expiratory wheezes on examination. The patient was admitted to an outside hospital two weeks ago and was diagnosed with asthma exacerbation complicated by pneumonia. Chest-X ray at outside hospital was significant for lobar infiltrates, and the patient was subsequently discharged on a course of antibiotics and prednisone. Patient continued to have sub-sternal chest pain and started experiencing new onset right lower extremity neuropathic symptoms without strength deficit or foot drop. On arrival in the Emergency Department the patient was tachycardic with EKG showing sinus tachycardia, troponin of 971 and BNP 1238. The WBC count was significant for absolute eosinophil count of 67%.

Figure. Multiple diagnostic modalities demonstrating eosinophilic myocarditis. A, ECG shows T-waves inversion lead 1, AVL, V3–V6, and ST depressions in V3–V6. B, Echocardiogram revealed normal left ventricle cavity size and predominant hypertrophy of the apaxes. C, Four-chamber echocardiogram showing apical hypertrophy and focal basal hypertrophy of the septum. D, Subendocardial thrombus along the apical interoseptal myocardium. E, Magnetic resonance imaging showed myocardial edema that is predominantly involving the mid to apical septal and interoseptal myocardial segments. F, Endomyocardium biopsy that shows patchy interstitial eosinophils (arrow pointing).

CT Angiogram chest showed bilateral consolidation compatible with ongoing pneumonia. Cardiology, pulmonology, and rheumatology were consulted for concerns for NSTEMI and eosinophilic pneumonia. ANCA battery, hypersensitivity pneumonitis evaluation, ANA workup were negative. IgE levels were 2605, TTE performed showed LV EF mildly reduced 50-54%, RA and RV dilation, trace pericardial effusion. CT abdomen noted to have colitis and duodenitis. Cardiac MRI showed evidence of myopericarditis, depressed LV function of 45% and mild RV dysfunction with no evidence of intracardiac thrombus.

Given patient’s cumulative multiorgan manifestations- including acute myocarditis, lung involvement, gastric involvement, questionable neurological involvement he was diagnosed with eosinophilic granulomatosis polyangiitis without definitive tissue biopsy. Patient was started on IV Solumedrol 1g x 3 days and was subsequently started on cytoxan IV 15mg/kg. Rheumatology recommended starting Mepolizumab 300mg SQ.

Conclusion

Patients suffering from refractory asthma and with high eosinophil counts should be alert to the possibility of having EGPA. EGPA associated eosinophilic myocarditis occurs rarely yet can be fatal if left untreated hence patients presenting with cardiac manifestations and eosinophilia should promptly be started on systemic corticosteroids.