An Unusual Case of Loffler endomyocarditis of the aortic value

Nisar A. Tramboo, K. Iqbal, B.A. Naikoo, Mehboob Ali Dar, Khalid Mohiudin, Manzoor Ali Andrabi

Abstract:

Idiopathic hypereosinophilic syndrome is a rare systemic disorder with an unexplained elevated eosinophil count. Loffler endomyocarditis is hypereosinophilic syndrome with endocardial fibrosis and restrictive cardiomyopathy. We describe a previously healthy 28 year male with Loffler endomyocarditis with restrictive cardiomyopathy involving right sided heart and moderate aortic regurgitation due to valve fibrosis and fibrotic vegetation of the aortic valve.

INTRODUCTION

Idiopathic hypereosinophilic syndrome is a leukoproliferative disorder, marked by sustained overproduction of eosinophils and its marked predilection to damage specific organs, including the heart. Frequently there is endocardial fibrosis with superimposed mural thrombus, resulting in restrictive cardiomyopathy described as Loffler endomyocarditis. The atrioventricular valves are frequently involved leading to atroventricular regurgitation. Previously there has been one case report of combined aortic and mitral valve involvement of Loffler endomyocarditis that was treated with bivalvular replacement and other case of Loffler endomyocarditis complicated by peripheral thromboembolism and severe aortic regurgitation due to valve fibrosis and fibrotic regurgitation on the aortic valve. We describe an unusual case of Loffler endocarditis with endomyocardial fibrosis of aortic valve with moderate aortic regurgitation. To our knowledge this is the third case only of Loffler endomyocarditis in which aortic valve is involved.

Material and Methods (report of a case)

A 28 year old male with unremarkable medical history in the past presented with symptoms of fever and right heart failure of twenty five days duration. The symptoms got relieved by salt restriction and diuretics over one week period. The patient also gave history of low grade fever of 100-101°F of the same duration more during evenings and associated with chills. There was also history of precordial pain intermittent type of the same duration and had no relation with exhaustion. Patient also complained of breathlessness with ordinary work. Clinical examination revealed general physical asthenic habitus with dark complexion pigmentation. His pulse rate was 80 beats/min regular and fair in volume. Brachial artery blood pressure of 110/70 mmHg, jugular venous pressure was 16 cm of H2O with prominent X and Y descent. Kusmaul’s sign was positive. There was no pedal edema. Chest examination revealed bilateral vesicular breath sounds with decreased intensity at bases. Cardiovascular examination showed apex beat at 5th intercostal space of hyperdynamic type. There was no thrill or any palpable sound. S1 and S2 were heard. P2 was of normal intensity, with right ventricular S3. Pansystolic murmur at tricuspid area which increased with inspiration. In addition there was early diastolic murmur at aortic area. He had no pericardial rub. Abdominal examination revealed hepatosplenomegaly with ascites. CNS examination was normal. Investigations done revealed Hb - 12.2 gm%, TLC - 53.69, DLC - P2L3E095, repeat TLC - 64.36, DLC - P1L4E095, ESR - 22mm/1 hr, platelet count - 1.19 lac, repeat 93000/cumm, normal urine exam. Liver and kidney function were normal. CkMB isoenzyme 29.0gm/lit. Hepatitis serology was negative. Coagulation and collagen profile were negative, blood culture was sterile and tuberculosis profile was negative. Stool for ova and cysts was negative. Bone marrow examination revealed hypercellular marrow with marked increase in eosinophils and its precursors.

Keywords: aortic valve, cardiomyopathy, endomyocarditis
with adequate iron stores, consistent with hypercellular marrow.

Ultrasound abdomen revealed free fluid in abdominal cavity and hepatosplenomegaly. X-ray chest revealed mild cardiomegaly with prominent right heart border, there was no PAH or PVI. ECG showed sinus rhythm, normal axis, negative p waves in lead I and AVL Q in and AVL and RSR pattern in V1.

Transthoracic echo revealed AO-3.0, LA 3.5, LV<5.0/3.2, EF 64%, RV - 3.5cm, RA and RV were dilated. Thickened tethered septal leaflet of tricuspid valve with thickened RV and LV apices. RV apex was obliterated (Fig. 1), AML flutters were present, pulmonary valve was normal, aortic valve was thickened. Colour doppler revealed severe low pressure tricuspid regurgitation with right ventricular systolic pressure of 35mmHg. Mitral value doppler study was normal and aortic valve colour doppler revealed aortic regurgitation of moderate severity (Fig. 2).

**Discussion**

In 1936 Loffler described two patients with progressive cardiac failure, eosinophilia and murmurs of mitral regurgitation. At autopsy, the patients had extensive fibrotic thickening of the mural endocardium of both right and left ventricles with superimposed thrombus in the left ventricle. The mural endocardial fibrosis and thrombosis were later described by Fauci et al to be virtually limited to the inflow tracts of the cardiac ventricles, more frequently involving the ventricular aspects of the posterior mitral and tricuspid valve leaflets. Hypereosinophilic syndrome is defined by unexplained eosinophilia (an eosinophil count >1500/L)(1), without evidence of parasitic, allergic or neoplastic cause for more than 6 months in the presence of end organ involvement. Eosinophil mediated heart damage evolves through three stages. The first state is an acute necrotic followed by thrombotic stage, which leads into a fibrotic stage clinical findings in the first stage can be absent with normal echocardiography and angiography. The second stage is characterized by the formation of thrombi along the damaged endocardium of either or both ventricles and occasionally the atrium, the outflow tracts near the aortic and pulmonary values are almost always spared, more commonly, a thrombus will form on the atrioventricular value leaflets. Finally the fibrotic stage, progressives scarring of the endomyocardium may lead to entrapment of chordae tendineae, causing mitral and or tricuspid cardiomyopathy. Two dimensional echocardiography is valuable in detecting. Intracardiac thrombi and thickening of posterior mitral valve leaflet and its attachement to the posterior wall. Cardiac catheterization may show increase in the right and left ventricular end diastolic pressures and valvular incompetence; it may also show apical obliteration or irregularities.

Hypereosinophilic syndrome patients often present at the later thrombotic and fibrotic stages with complaints of dyspnoea or chest pain, signs of left and/or right ventricular congestive heart failure, murmurs of mitral regurgitation, cardiomegaly and/or ‘T’ wave inversion on electrocardiography. Those who develop thrombosis may present with thromboembolism.

The case under discussion was a 28 years male who presented to us with a recent onset right heart failure and with echo documented restrictive cardiomyopathy involving right heart and endomyocardial fibrosis of the aortic valve with moderate aortic regurgitation. The patient had marked eosinophilia confirmed on PBF and bone marrow biopsy. The patient was managed with anti-failure treatment and prednisolone (1mg/kg), during hospital stay he showed objective improvement, however there was no change in eosinophil count, the patient was lost to follow up.

To our knowledge this is the third case in the literature in which Loffler endomyocarditis involves the aortic valve.

**REFERENCES:**