Case Report

Recurrent Loffler’s Endomyocarditis: Clinical and Operative Challenges

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Abstract

This patient had primary Hypereosinophilic endomyocarditis “Loffler’s Endomyocarditis” complicated by intraventricular thrombosis, mitral and tricuspid valve regurgitation, and associated with heparin induced thrombocytopenia syndrome. He underwent valve surgery twice for recurrence of Left ventricular thrombosis and valve dysfunction.

This rare disease is characterized by peripheral hypereosinophilia and involves the heart in 75% of the cases. The disease carries poor prognosis due to endomyocardial fibrosis, ventricular and atrioventricular valve dysfunction and tendency to develop intraventricular thrombi.

CASE PRESENTATION

This 40 years old male patient presented with history of night sweats and weight loss for six month followed by persistent dry cough and progressive dyspnea. He had a history of Bilharziasis which was treated 20 years ago.

Chest examination was clear while cardiovascular examination revealed pan systolic murmur over the apex. ECG showed normal sinus rhythm, and Chest X-ray showed pulmonary vascular congestion with bilateral small pleural effusion consistent with congestive heart failure. Blood count revealed a white blood cell count of 24,600/mm³ with hypereosinophilia (4280/mm³). Echocardiography showed a large thrombus in the left ventricle (LV) (Figure 1 a-c) involving the endocardium of the ventricular septum, anterolateral wall and extending to posterior basal wall almost protruding through the LV outflow tract (Figure 2), but with no significant signs of obstruction. There was severe mitral regurgitation (Figure 3) due to involvement of the anterior mitral leaflet, and also moderately severe tricuspid regurgitation with a dilated tricuspid annulus and pulmonary artery systolic pressure of 65mmHg. The diagnose of primary Hypereosinophilic (HES) endomyocarditis (Loffler endomyocarditis) was highly suspected. Abdominal Ultrasound showed splenomegaly with heterogeneous parenchyma. Bone marrow and splenic biopsies showed a heavy eosinophilic infiltration with no evidence of leukemia or parasitic infection (Figure 4 a,b). He received anticoagulation and intensive steroid therapy. During therapy, he developed acute reduction of platelet count more than 50% and Heparin-Induced Thrombocytopenia (HIT) test was positive. Surgery was planned because of valve involvement and persistent heart failure [1-6].

Operative management

Because of the HIT syndrome, i.v Lepirudine for anticoagulation during bypass [10] was used and monitored by measuring activated partial thromboplastin time “APTT” (Ecarin clotting time [7], which is more reliable to measure the Lepirudine thrombin inhibiting effect, was not available). Our target APTT was >180sec and was monitored every 15min. Via a left atriotomy, a good access to the mitral valve and LV thrombus was achieved. Total excision of the mass and endomyocardial decortications was achieved. The mass involved the anterior mitral leaflet and subvalvular apparatus as well as the posterior mitral leaflet. The whole mitral valve apparatus was excised and replaced using a bioprosthesis. The aortic valve was inspected and there was no involvement of its leaflets, the LV outflow tract was cleaned and any residual thrombus was removed. Via right atriotomy inspection of the tricuspid valve revealed only dilated annulus (Functional TR). The regurgitation was corrected using a tricuspid anuloplasty ring. The patient tolerated the procedure very well and came off bypass with minimal inotropic support.

Postoperatively, the patient was kept on warfarin, steroids, hydroxyurea and Gleevic (Imatinib Mesylate). He was discharged home in good condition on the 10th postoperative day for regular follow up. Echocardiography study at six month showed no abnormalities.

The patient presented to the clinic with severe heart failure symptoms 11 months after surgery. Checking his compliance with medications, it was found that he stopped the hydroxyurea drug for the last four months. Repeated echo showed recurrent LV mass infiltrating the mitral bioprosthesis with partial obstruction of the mitral inflow (Figure 2). Because the patient was known HIT positive, the test was repeated and found to be negative. Redo surgery was carried out through a median sternotomy, dissection of tough pericardial adhesions, aortic and bicaval cannulation were done. Heparin was used as anticoagulant and CPB was instituted. The mitral valve was accessed through transeptal approach, explanted with the infiltrating thrombi, and the LV cavity was cleaned. A Mitral bioprosthesis was implanted.

The patient’s recovery was uneventful and was discharged home after one week. Follow up echo study after one year showed no abnormalities.

**DISCUSSION**

In 1936 William Loffler described hypereosinophilia and progressive heart failure [1-3]. It is characterized by acute necrotic stage, followed by thrombosis and finally fibrosis leading to restrictive cardiomyopathy [4].

Treatment options in this disease are limited. Steroids is the first line treatment in this disease followed by cytotoxic drugs (hydroxyurea), tyrosine kinase inhibitors, monoclonal antibody therapy and even bone marrow transplant [3].

Loffler endomyocarditis is associated with poor prognosis; however endomyocardial decortications and bioprosthetic valve replacement is associated with a 5yr survival rate of 72% [2].

The tendency to develop thrombosis in our patient due to HES was complicated by HIT syndrome. HIT is an immunological disorder seen in 1-3% of patients receiving unfractionated heparin resulting in low platelet count and activation of platelet aggregation with a high risk of thrombosis and mortality rate of 28%. It is seen in 0.7%-1.9% of cardiac surgery patients [5].

Laboratory tests can be used to support the diagnosis of immune HIT. However, the negative predictive value of these tests is generally < 50%, so that a negative result does not exclude...
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the diagnosis. Furthermore, 50% or more of patients exposed to heparin without the development of HIT develop detectable antibodies by ELISA[5,9,10]. This rare combination of HES and HIT represented a surgical challenge. The management entitled endomyocardial decortications with valve repair or replacement together with HIT management.

Moraes and colleagues reported 83 patients who underwent endomyocardial decortications and atroioventricular valve repair or replacement with a follow up ranging between 2 month and 17 years. Actuarial survival at 17 years in their group, including operative mortality, was 55% with a recurrence rate requiring re-intervention of 8.8% [6-8].

In general, valve repair is not recommended in these cases due to high failure rate.

The case we are reporting shows all the complications possible in case of HES [9,10], including HIT and recurrence of intracavitary mass due to interruption of appropriate medical treatment. It emphasizes that aggressive surgical treatment in case of recurring intracavitary thrombosis can be successful.

REFERENCES