

Cardiac Mucormycosis with T-cell Immunodeficiency

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We present a case of a 2 year old apparently healthy child who presented with fever and mass on the mitral valve. Excision histopathology of the mass revealed mucormycosis. After 4 months, she had CNS embolisation with recurrence of cardiac lesion when investigations revealed associated T-cell immunodeficiency.

Keywords: *Cardiac mucormycosis, Immunodeficiency, Mitral valve, T-cell defect.*

Mucormycosis is a rare, progressive, systemic necrotizing fungal infection rarely affecting a normal human host. Most cases of cardiac mucormycosis described in literature have been diagnosed on autopsies(1). We describe a case of cardiac mucormycosis in a child with structurally normal heart who was also detected to have immunodeficiency. This case illustrates the extensive nature of organ involvement in mucormycosis and its resistance to available treatment.

CASE REPORT

A 23-month old apparently healthy girl presented with a one month history of high grade fever and one episode of abnormal movements of left side of the body. On examination, she weighed 9.5kg, was febrile and had tachycardia. All peripheral pulses were well felt. Cardiovascular examination revealed normal heart sounds and a short systolic murmur at the apex. Respiratory, abdominal and central nervous system examination were normal. Hemoglobin was 10.4 g/dL, total count 7300/mm³ (20% polymorphs and 76% lymphocytes) and ESR

45mm/hour. Liver and renal function tests were normal. Random blood sugar was 80 mg/dL and HIV serology was negative. Urine routine microscopy revealed 6-8 RBCs/hpf. Chest X-ray and electrocardiogram were normal. Echocardiogram revealed a large pedunculated mass on the superior surface of anterior mitral leaflet with an additional small mass on the superior surface of posterior mitral leaflet and mild mitral regurgitation. Endocarditis and myxoma were considered to be the likely possibilities. A day later, the child underwent excision of the masses with mitral valve repair. During surgery, small satellite lesions were noted on mitral valve chordae. Histopathological examination of the excised mass revealed it to be a fungal vegetation consistent with mucormycosis. The blood and urine cultures sent prior to surgery were negative. The child was treated with parenteral amphotericin B (45 mg/kg cumulative dose) with close monitoring of liver and renal function tests. She was discharged on oral fluconazole (5 mg/kg/day) in an afebrile state. Echocardiogram at the time of discharge did not reveal any vegetations.

The child was brought back four months later

with altered sensorium, generalized seizures, weakness of all four limbs and inability to speak and swallow for 24 hours. On examination, she was drowsy and had persistent focal seizures. The tone was increased in the right lower limb with increased deep tendon reflexes. She had grade 2/5 power in left upper and lower limbs. Cardiovascular examination revealed a systolic murmur at the apex with normal heart sounds. CT scan head showed left basal ganglion and left parieto-occipital infarcts with old right fronto-parietal infarcts. The right internal carotid artery was noted to be blocked with thrombus on a CT angiogram. Echocardiography revealed moderate mitral regurgitation with a 10 mm vegetation on the anterior mitral leaflet (**Fig. 1**). Serial blood cultures grew *Candida*-non albicans type. Lymphocyte enumeration study was done which showed decreased CD3+/CD45+ (T-cells), CD3+/CD4+ (T-helper cells) and absolute CD4+ lymphocyte count. Intravenous amphotericin B was restarted. Oral fluconazole was continued. After four weeks of therapy the neurologic status gradually improved, however left sided hemiplegia persisted. The mitral regurgitation had worsened on echocardiography. After 12 weeks of parenteral treatment, the child was discharged on oral antifungals.

DISCUSSION

Mucormycosis is an inclusive term for progressive disseminated infections caused by phycomycetes, which are usually limited in their virulence but can be highly invasive in certain conditions. The most often affected sites are lungs, heart, liver, spleen and kidneys. Dissemination has been most extensive in cases with endocardial involvement(2). It can occur *de novo*(1,3,4) or in association with cardiovascular surgery(5). Mucormycosis involving native hearts have usually occurred in immunodeficient patients with few exceptions(4). Hence, in this case, we were prompted to evaluate for immunodeficiency. Our case is extremely rare as affliction by mucormycosis of native heart valve in a two year old remains undescribed(3-7).

The major controversial issue in cardiac mucormycosis is treatment. Surgical excision followed by amphotericin appears to be the most accepted

strategy as described by Sanchez-Recalde, *et al.*(5). Administration of antifungal therapy for atleast one week before surgery is recommended to reduce the fungal burden whenever feasible. This is followed by 5 weeks course of parenteral antifungals. Amphotericin B can be ineffective in patients whose disease is detected late or who have disseminated disease(8). Liposomal amphotericin B is considered as the drug of choice as it allows high intra-macrophage concentration allowing high concentration of the drug within the vegetation(9). Combination therapy using lipid based amphotericin with an echinocandin or with an azole (largely itraconazole or posaconazole) or with all three is preferred(10). In our case, on initial presentation, chances of embolisation from a pedunculated mass prompted us for direct surgical intervention and the child did not receive any preoperative antifungal coverage. Combined approach of surgical and 12 weeks of post operative amphotericin was not sufficient. On readmission, medical treatment was thought to be the best possible therapy in view of the disseminated nature and difficulty in demarcating the extent of cardiac involvement.

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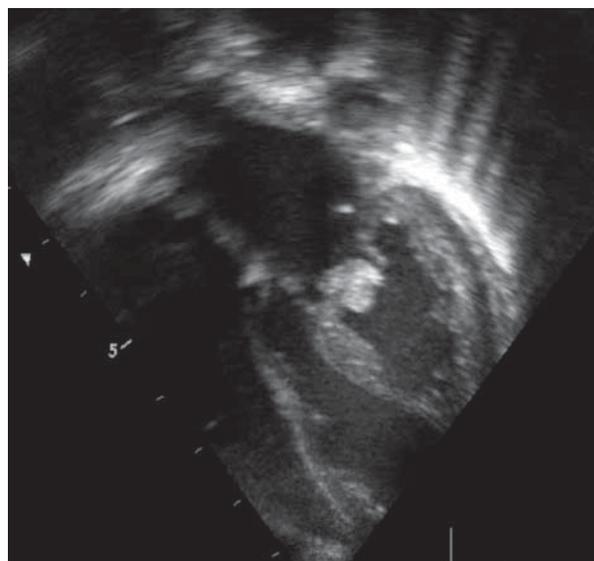


FIG.1 2D echocardiogram performed 4 months after surgery showing a mass attached to anterior mitral leaflet with respective chordae thickened in apical four chamber view.

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REFERENCES

1. Basti A, Taylor S, Tschopp M, Sztajzel J. Fatal fulminant myocarditis caused by disseminated mucormycosis. *Heart* 2004; 90: e60.
2. Scully RE, Mark EJ, McNeely WF, Ebell SH, Philips LD, Ellender SM. Weekly clinicopathological exercises. *NEJM* 1999; 341: 265-273.
3. Mehta NN, Romanelli J, St. John Sutton MG. Native aortic valve vegetative endocarditis with *Cunninghamella*. *Eur J Echocardiogr* 2004; 5: 156-158.
4. Chandrasekar P, Rao SR, Raman ML, Vaidyanathan PR, Muralidharan S. Mucormycosis of the heart presenting as paraparesis. *Indian Heart J* 1998; 50: 75-76.
5. Sanchez-Recalde A, Merino JL, Dominquez F, Mate I, Larrea JL, Sobrino JA. Successful treatment of prosthetic aortic valve mucormycosis. *Chest* 1999; 116: 1818-1820.
6. Choudhry R, Venugopal P, Chopra P. Prosthetic mitral valve mucormycosis caused by mucor species. *Int J Cardiol* 1987; 17: 333-335.
7. Callard GM, Wright CB, Wray RC, Minor GR. False aneurysm due to mucor following repair of a coarctation with a dacron prosthesis. *J Thorac Cardiovasc Surg* 1971; 61: 181-185.
8. Sugar AM. Agents of mucormycosis and related species. *In: Mandell GL, Bennett JE, Dolin R, eds. Mandell, Douglas and Bennett's Principles and Practice of Infectious Diseases. 5th ed. Philadelphia: Churchill Livingstone; 2000. p. 2685-2695.*
9. Ellis M. Amphotericin B preparations: a maximum tolerated dose in severe fungal infections? *Transpl Infect Dis* 2000; 2: 51-61.
10. Spellberg B, Edwards J, Ibrahim A. Novel perspectives on mucormycosis: Pathophysiology, presentation and management. *Clin Microbiol Rev* 2005; 18: 556-569.