Fungal Endocarditis in an Alport’s Syndrome Patient with Chronic Kidney Disease on Regular Hemodialysis

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ABSTRACT

Introduction: Fungal Endocarditis is rare and fatal. Most prevalent in immunosuppressed and Intravenous Drug Abusers. Candida and Aspergillus species are most common etiologic Fungi.

Case presentation: A Seventeen-year-old male with known Hypertensive, Seizures and Alport Syndrome of Stage V Chronic Kidney Disease, admitted with history of fever and chills for 2 weeks. On regular hemodialysis via right Internal Jugular Vein Pencath, inserted 6 months ago. Clinical examination was unremarkable except tachycardia and hypotension. Serial blood cultures were negative for any growth. But 2D Echocardiography confirms the presence of mobile vegetative mass attached to Tricuspid Valve (TV), which extends and then obstructs Right Ventricular Outlet Tract (RVOT). With worsening symptoms and failed initial management, he was admitted first time and underwent removal of Pencath, TV Vegetectomy and then Pericardial patch augmentation of Septal Tricuspid Leaflet and Alferi type of TV repair. Both Pencath tip and excised mass were sent for Histopathological Examination (HPE), confirms Fungal infection with Magnusiomyces Capitatus, belongs to Blastoschizomyces capitatus. Responds to Inj. Loposomal Amhotericin B and Voroconazole. But within few weeks, he developed Severe Tricuspid Regurgitation and Right heart failure with gross Ascitis. Then readmitted for second time and underwent immediate symptomatic relief by Ascitic tapping followed by TV Replacement with 31 mm of St. JUDE Mechanical Valve. Perioperative management was challenging and stabilized gradually and involves a multispecialty team approach.

Discussion: Fungal endocarditis is a serious condition. Combined aggressive Medical and Surgical therapy will have better outcome.

Key Words: Antifungal Agents, Fungal Endocarditis, Right Heart Failure, Tricuspid Valve Replacement, Ascitic tapping, Multispecialty team approach

INTRODUCTION

Fungal endocarditis (FE) remains the rare most serious form of infective endocarditis, with a high mortality rate of about 50%.¹²³ It is highly challenging to identify the source, establish a diagnosis and to apply specific treatment. This is because of its high mortality rate and complexity of FE will lead to lack of prospective randomized clinical trials for FE management.

CASE REPORT

We report a case of Fungal Endocarditis in a Seventeen-year-old male patient. He suffers from Hypertension, Seizure disorder and Alport’s Syndrome with Chronic Glomerulonephritis with Stage 5D chronic kidney disease. He inherited Alport’s Syndrome from his mother. She also had CKD, both were on regular hemodialysis. His female sibling was free from Alport’s Syndrome. Alport’s Syndrome - X linked Ge-
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Genetic disorder mainly affects kidney. He undergoes hemodialysis via Permcath, which was placed 6 months ago at Right Internal Jugular vein. He was admitted twice with intractable clinical course for Fungal Endocarditis management and stays in the hospital for nearly about 92 days.

First Admission: He was clinically presented with history of fever, chills and tachycardia. All of his investigations were unremarkable except-Anemia (5.5 gms/dl), Thrombocytopenia (17000 cells /cumm) and 2D Echocardiography confirms presence of a mobile mass attached to Tricuspid Valve (TV) leaflet as shown in Figure 1. Presence of Fever with chills and tachycardia, rapid growth of mass and presence of indwelling catheter confirms Infective Vegetative mass and hence Intra Cardiac mass as differential diagnosis was ruled out. Serial blood cultures were revealed no growth, probably because of prior of medications. He failed to respond initial use of broad-spectrum antibiotics and antifungal treatments on outpatient basis. As constitutional symptoms persist and increased Right Ventricular Outlet Tract obstruction by TV vegetative mass, with no option left out relatives were counselled, consented and admitted for high-risk TV mass removal surgery. Prior to surgery, CT Thorax was done which showed extension of TV mass as in Figure 2. Also, he receives Six units of Leucodepleted Packed Red Blood Cells and Four units of Single Donor Platelet during periprocedural period for correction of Anemia and Thrombocytopenia. Immediately after admission, Permcath was removed as it may be the only source of infection. Then Left Femoral HD catheter was placed for regular in-hospital temporary hemodialysis. Under General Anesthesia with Cardio Pulmonary Bypass and Mechanical Ventilation support, TV mass (Size - 3 X 4 x 5 cms) was removed by TV Vegetectomy and then Pericardial patch augmentation of Septal Tricuspid Leaflet and Alferi type Tricuspid Valve repair (by TV edge to edge stich) was done. Both Permcath tip and TV mass were sent for histopathological examination (HPE) analysis. Both confirms Fungal Endocarditis (Fungal IE) as shown in Figure 3 - Magnusiomyces Capitatus, belongs to Blastoschizomyces capitatus. Figure 3 also shows gross anatomy of the excised mass. Along with regular medications. he was treated with Inj. Loposomal Amhotericin B, Tab. Voriconazole and other supportive measures. Then gradually weaned from Mechanical Ventilation support and extubated. Before first discharge, temporary Left Femoral Hemodialysis catheter was removed and new Left Brachiocephalic Fistulae was created for future permanent hemodialysis. Prior to first discharge 2D Echo shows Moderate TR as shown in Figure 4.

Second Admission: Within few weeks, he was readmitted with worsened breathing difficulty and gross ascites. 2D Echo shows Dilated RA, RV, Severe TR and RV dysfunction as in Figure 5. Immediately he underwent therapeutic Ascitic fluid aspiration and aspirated 3.5 liters of Ascitic fluid to get immediate symptomatic relief. As TV destruction

![Figure 1: 1st Admission Pre Surgery 2D Echo stills of both TTE and TEE](image1)

![Figure 2: CT Stills of Thorax](image2)

![Figure 3: Gross macroscopic and Microscopic Pictures of Tricuspid Valve Vegetative mass](image3)

![Figure 4: 1st Admission Post Surgery TTE 2D Echo stills.](image4)
continued, Tricuspid Regurgitation worsens and so Tricuspid Valve Repair (TVR) was advised. Again relatives were counselled, consented and admitted for high-risk 2nd Surgery. Again, under General Anesthesia with Cardio Pulmonary Bypass and Mechanical Ventilation support - underwent successful TV Replacement surgery with 31 mm of St. JUDE Mechanical Valve. He had delayed weaning process from Mechanical Ventilation support due to intractable Seizures, treated with IV Sodium Valproate. We also encountered and managed some of the other complications that include Valproate induced Pancreatitis - treated conservatively, Left Femoral Artery Pseudoaneurysm and Hematoma - managed with direct Thrombin injection and Pseudoaneurysmal repair & large Left thigh hematoma evacuation and also had anteriorly popped outed Sternal wire, which was later removed directly. He was then gradually stabilized. Throughout his hospitalizations, he underwent regular hemodialysis initially via temporary Left Femoral Hemodialysis catheter and later with permanent Left Brachiocephalic Fistulae. At 2nd discharge, 2D Echo has showed definitive improvement as shown in Figure 6 with mild TR and without RV dysfunction. Multi-Specialty consultations was done including Diet, Psychiatry and Genetic counselling. Plan for Renal Transplantation after stabilization at later date.

**DISCUSSION**

Fungal endocarditis (FE) is fatal, usually being diagnosed postmortem.\(^1\) FE is most prevalent in patients who are immunocompromised, intravenous drug abusers, prolonged use of antibiotics, parenteral nutrition or any intracardiac devices.\(^1,2\) The diagnosis of FE is challenging due to its slow growth.\(^1,2\) Neither any specific tests nor any diagnostic criteria were available for Fungal IE.\(^2\)

High mortality rate and complexity of FE may lead to lack of prospective randomized clinical trials for FE management.\(^2\) Current guidelines recommend initial or induction therapy with Amphotericin B (AMB) with or without Flucytosine combined with surgical removal of vegetations, followed by chronic suppressive therapy with oral fluconazole.\(^1,2\) Voriconazole is for both first line for induction and long-term suppression therapy for treating Aspergillus endocarditis.\(^2\) Duration of antifungal therapy is usually 6-8 weeks depending upon clinical improvement.\(^2\) Combined treatment

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**Table 1: Indications and Timing of Surgery in Fungal Endocarditis**

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<tr>
<th>Indications for surgery</th>
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<tr>
<td>Class I</td>
<td>Valve dysfunction resulting in symptoms of heart failure</td>
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<tr>
<td>Class I</td>
<td>Left-sided IE by S. aureus, fungal, or other highly resistant microorganisms</td>
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<tr>
<td>Class I</td>
<td>Complicated by CHB, annular / aortic abscess, or destructive penetrating lesions</td>
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<tr>
<td>Class I</td>
<td>Persistent infection 5-7 days after initiation of appropriate antibiotic therapy</td>
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<tr>
<td>Class Ia</td>
<td>Prosthetic valve endocarditis with relapsing infection</td>
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<td>Class Ia</td>
<td>Recurrent emboli and persistent vegetations despite appropriate antibiotic therapy</td>
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<tr>
<td>Class I</td>
<td>Patients should be on appropriate antibiotic therapy at the time of surgery</td>
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<tr>
<td>Class Ia</td>
<td>Once a patient is on an appropriate antibiotic regimen, further delay of surgery is unlikely to be beneficial</td>
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appears to be superior to monotherapy, before the onset of valve destruction, fatal embolic strokes or chordae rupture causing valvular insufficiency.\textsuperscript{1,4} In the presence of Fungal Endocarditis, Indications and Timing of surgery is very important. Indications and Timing of Surgery in the presence of Fungal Endocarditis were listed in Table 1 - a Consensus on Surgical Treatment of Infective Endocarditis by American Association for Thoracic Surgery Guidelines.\textsuperscript{5}

**CONCLUSIONS**

Fungal endocarditis (FE) is a serious health condition. Early diagnosis and intensive managements are essential. Combined Medical and Surgical therapy will have better outcome. Consensus on FE management is evolving.

**Learning Points**

Aggressive and early combined Medical and Surgical therapy for FE is essential.

Multi-Specialty approach is often required.

Dietary Consultation and Psychiatry Counselling are equally important.

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**REFERENCES**