International Journal of Current Research and Review
DOI: http://dx.doi.org/10.31782/IJCRR.2022.14809

Review Article

IJCRR
Section: Healthcare
ISI Impact Factor (2021-22): 2.176
IC Value (2020): 91.47
SJIF (2020) = 7.893

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 2weeks. On regular hemodialysis via right Internal Jugular Vein Permcath, 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 removable of Permcath, TV Vegetectomyand then Pericardial patch augmentation of Septal Tricuspid Leaflet and Alferi type of TV repair. Both Permcath tip and excised mass were sent for Histopathological Examination (HPE), confirms Fungal infection with Magnusiomyces Capitatus, belongs to Blastoschizomyces-capitatus. Responds to Inj. LoposomalAmhotericin 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 endocarditisis 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%.[1,2,3] 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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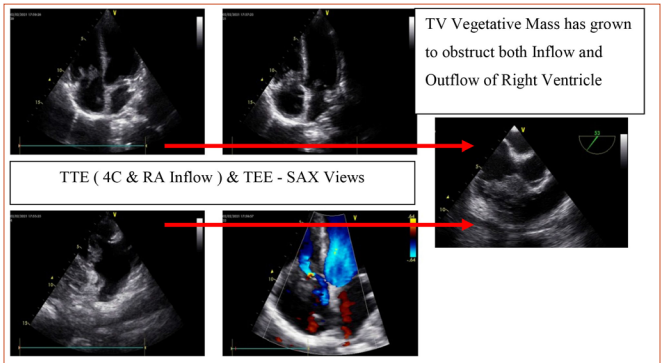
ISSN: 2231-2196 (Print) **ISSN:** 0975-5241 (Online)

Received: 10.01.2022 **Revised:** 08.02.2022 **Accepted:** 12.03.2022 **Published:** 19.04.2022

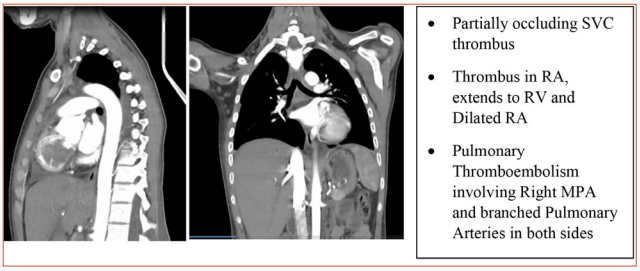
netic disorder mainly affects kidney. He undergoes hemodialysis via Permcath, which was placed 6 months ago at Right Internal Jugular vein. He was admitted twice with intractable clinical course for Fungal Endocarditis management and stays in the hospital for nearly about 92 days.

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. Voroconazole 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 Fiastulae was created for future permanent hemodialysis. Prior to first discharge 2D Echo shows Moderate TR as shown in **Figure 4**.



TTE = Trans Thoracic Echocardiography : TEE = Trans Esophageal Echocardiography : RA = Right Atrium : TEE - SAX Views = Trans Esophageal Echocardiography - Short Axis View : TV = Tricuspid Valve : 2D Echo = 2 Dimensional Echocardiography

Figure 1: 1st Admission Pre Surgery 2D Echo stills of both TTE and TEE



CT = Computaeized Tomography: SVC = Superior Vena Cava : RA = Right Atrium : RV = Right Ventricle : MPA = Main Pulmonary Artery: 2D Echo = 2 Dimensional Echocardiography

Figure 2: CT Stills of Thorax

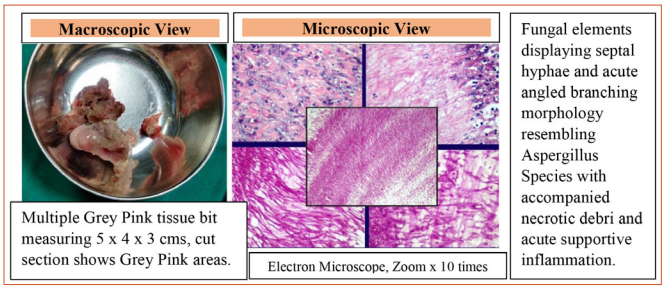
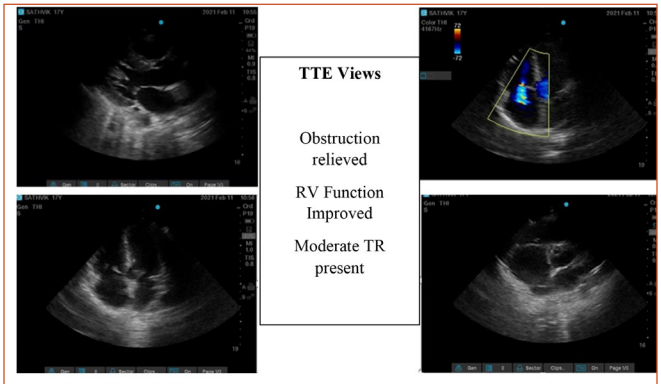


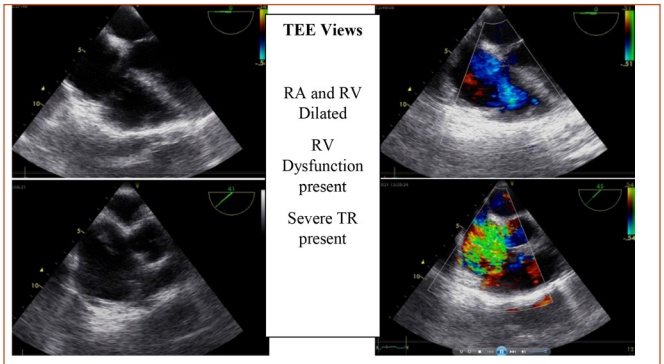
Figure 3: Gross macroscopic and Microscopic Pictures of Tricuspid Valve Vegetative mass

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



TTE = Trans Thoracic Echocardiography: 2D Echo = 2-DimensionalEchocardiography:RV = Right Ventricle:TR = Tricuspid Regurgitation

Figure 4: 1st Admission Post Surgery TTE 2D Echo stills.



TEE = Trans Esophageal Echocardiography: RA = Right Atrium: RV = Right Ventricle: TR = Tricuspid Regurgitation

Figure 5: 2nd Admission Pre Surgery TEE 2D Echo stills

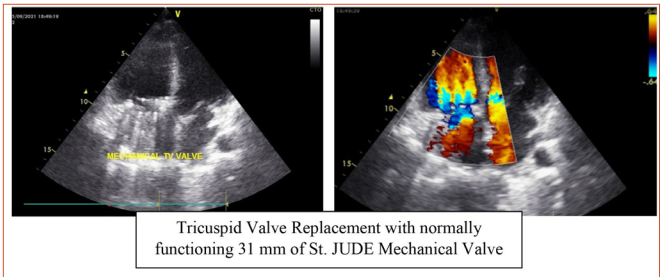


Figure 6: 2nd Admission Post Surgery TTE 2D Echo stills

TTE = Trans Thoracic Echocardiography: 2D Echo = 2-Dimensional Echocardiography

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 Fiastulae. 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

Table 1: Indications and Timing of Surgery in Fungal Endocarditis5

Indications for surgery		
Class I	❖	Valve dysfunction resulting in symptoms of heart failure
Class I	❖	Left-sided IE by S. aureus, fungal, or other highly resistant microorganisms
Class I	❖	Complicated by CHB, annular / aortic abscess, or destructive penetrating lesions
Class I	❖	Persistent infection 5-7 days after initiation of appropriate antibiotic therapy
Class IIa	❖	Prosthetic valve endocarditis with relapsing infection
Class IIa	❖	Recurrent emboli and persistent vegetations despite appropriate antibiotic therapy
Indications for surgery		
Class I	❖	Patients should be on appropriate antibiotic therapy at the time of surgery
Class IIa	❖	Once a patient is on an appropriate antibiotic regimen, further delay of surgery is unlikely to be beneficial

appears to be superior to monotherapy, before the onset of valve destruction, fatal embolic strokes or chordae rupture causing valvular insuffiency.[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.[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.

Acknowledgements

We acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. We are also grateful to authors / editors / publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

Conflict of interest: No conflict of interest

Source of funding:None

Authors contribution:

Nithin Bharadwaj contributes to complete conceptualization and data collection, Chandra Shekara Reddy creates complete manuscript preparation and editing, Arun Srinivas as corresponding author and provides complete guidance and proof correction of manuscript, Siddarth Kumar Chawath guides complete journal search and publication guidelines and Lakshminarayan Achar permits to access complete surgical notes and figures / photos.

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