Aortic root abscess with complete heart block in a patient with acute lymphoblastic leukaemia

Authors
Dr Harish M M¹, Dr Ramya B M², Dr J.V.Divatia³, Dr A.P.Kulkarni⁴, Dr Amol T Kothekar⁵
¹Chief Consultant Intensivist, ²Senior Resident, ³,⁴Professor, ⁵Assistant Professor
Department of Anaesthesiology, Critical Care Medicine and Pain, Tata Memorial Hospital, Mumbai, Maharashtra
Corresponding Author
Dr Harish M M
Chief Consultant Intensivist, Department of Anaesthesiology, Critical Care Medicine and Pain
Tata Memorial Hospital, Tata Memorial Hospital, Parel, Mumbai-400012

Abstract
Infective endocarditis involving left side of the heart is more common than the right side. Aortic valve involvement is relatively less common than the mitral valve and is usually seen in the setting of underlying pathologies like bicuspid or quadricuspid valve, rheumatic heart disease and prosthetic valve. Conduction abnormalities occur in 45% of patients with aortic vegetation with perivalvular abscess with an underlying bicuspid or quadricuspid valve. Endocarditis and aortic valve root abscess in absence of such pathologies is a very rare occurrence. We report here a rare case of aortic valve root abscess without any underlying structural abnormality with complete heart block in an Immunocompromised patient.

Case Report: A 19 yr male patient with B-cell acute lymphoblastic leukemia (ALL) who had received induction chemotherapy, presented to casualty with history of fever, cough and breathing difficulty since 2 days. The ECG showed premature ventricular escape beats. Subsequently ECG showed bradycardia and complete heart block leading to cardiac arrest. After successful resuscitation a transvenous pacemaker was inserted owing to persistent heart block. Artificial ventilation was continued. Persistent hypotension was treated by high dose adrenaline and vasopressin infusions. Transthoracic echocardiography showed aortic valve vegetations and perivalvular root abscess with moderate to severe aortic regurgitation. Patient remained hypotensive despite vasopressors and eventually expired.

Conclusion: Though aortic valve endocarditis is usually seen in patients with underlying conditions like bicuspid aortic valve or rheumatic valvular heart disease it may rarely be seen in patients without any of such underlying conditions and should be considered during evaluation of any patient presenting with features consistent with aortic valve endocarditis especially in the Immunocompromised patients.

Keywords: Aortic root abscess, complete heart block, acute lymphoblastic leukemia, Immunocompromised patients.

Introduction
Infective endocarditis is defined as an infection of endocardial surface of the heart. It may involve mural endocardium, single or multiple heart valves and septal defects [1]. Various conditions predisposing an individual for development of
endocarditis include congenital heart diseases (ventricular septal defect, persistent ductus arteriosus, tetralogy of Fallot, bicuspid aortic valves or aortic root dilatation as seen in Marfans syndrome), Rheumatic valvular heart diseases and degenerative heart disease (calcific aortic stenosis) [2]. In addition to these conditions prosthetic valves, intravenous drug users, nosocomial infections and fungal endocarditis in immunocompromised patients are also common causes of endocarditis [3]. Though knowledge of pre-existing cardiac conditions predisposing an individual for development of endocarditis may help in early diagnosis it should be kept in mind that infective endocarditis and aortic root abscess may develop in absence of any structural heart disease [4].

Aortic root abscess is rarely seen in patients with infective aortic valve endocarditis. It is a severe form of infective endocarditis presenting as a challenge to treating cardiologist. It may present as persistent fever, intractable cardiac failure, cardiac fistula, pseudo-aneurysm or varying degrees of heart block leading to hypotension and bradycardia [5]. Early diagnosis and surgical treatment of a perivalvular abscess are essential in preventing severe valvular destruction and improving the survival rate. Diagnosis is usually done by blood culture and echocardiography [6]. Conduction abnormalities that develop in patients with active infective endocarditis may represent extension of infection from the valve leaflets into the surrounding myocardium [7]. Extension of infection from the aortic valve into the septum can lead to significant conduction abnormalities, with bundle-branch blocks or complete heart block. The treatment consist of antibiotics, early reconstruction of aortic valve, valve replacement and cardiac pacing. Any delay in treatment is dangerous and may prove fatal [8].

We are reporting a rare case of aortic valve root abscess in an Immunocompromised individual without any underlying structural abnormality presenting with complete heart block.

**Case Report**

A 19 yr male patient with B - cell acute lymphoblastic leukemia who had received induction chemotherapy presented to casualty with history of fever cough and breathing difficulty since 2days. On clinical examination he was found to have irregular pulse (rate 110-120 beats/minute). Respiratory distress was present in the form of tachypnea (respiratory rate= 42/min ). On auscultation he had early diastolic murmur (Grade-III) heard in the aortic area. The ECG showed premature ventricular escape beats. He was hypotensive at the time of admission with a blood pressure of 98/62 mm of hg.

![Figure 1: Electrocardiograph showing premature ventricular beats with complete heart block (Left). Restoration of rhythm after pacing (Right).](image-url)

Patient was shifted to ICU. His investigations revealed normal hepatic and renal function tests, complete blood count showed thrombocytopenia and lymphocytosis and anemia (Hb- 5.9 gm) blood gas showed mild respiratory alkalosis. He was started on Non Invasive Ventilation (NIV).
The rhythm converted to sinus rhythm after NIV. Patient was started on broad spectrum antibiotics and transfused 2 units of packed cells. He remained on NIV for whole night and was haemodynamically stable. Next day early in the morning patient was complaining of dizziness. ECG was taken which shows bradycardia and complete heart block, within few minutes he had sudden cardiac arrest. He was resuscitated with ROSC (return of spontaneous circulation) of 1 min 30 seconds. ECG still showed persistent complete heart block therefore transavenous pacemaker was inserted through right internal jugular vein. The pacemaker settings were demand mode, HR-90, sensitivity-1mv, output-4v. He required high dose adrenaline and vasopressin infusions due to persistent hypotension. Mechanical ventilation was continued on assist volume controlled mode with high PEEP and 100% FiO₂. Transthoracic echocardiography was done, which showed aortic valve vegetations and perivalvular root abscess with moderate to severe aortic regurgitation.

**Figure 2**: 2D Echocardiography showing aortic valve vegetations with aortic root abscess.

After repeating blood cultures, antibiotics were changed to meropenem, caspofungin, and vancomycin. Considering the possibility of complicated malaria artemunate was added empirically. Cardiovascular surgeon’s opinion was taken and in view of unstable haemodynamics he advised continuation of medical line of management with transvenous pacing and further management after stabilisation (valve replacement). Repeated blood gases showed severe metabolic acidosis with high lactates, patient remained persistently hypoxic and hypotensive and expired in the evening.

**Discussion**

Aortic root abscess is one of the unusual complications of aortic valve endocarditis [9]. In the International Collaboration on Endocarditis Merged Database (ICE-MD) study, among 311 patients with aortic valve endocarditis, 22% had periannular abscesses [10]. Complete heart block occurs in 5–14% of cases of infective endocarditis. The right and non-corporal sinus of Valsalva lie close to the superior inter-ventricular septum and bundle of His. The infection of aortic root with abscess formation is associated with life-threatening conduction abnormalities because the infection spreads through the septum. This causes abnormalities which include bundle branch and atioventricular (AV) block [11]. Isolated aortic valve vegetations are less common in patients with structurally normal aortic valve, but can be present in bicuspid/quadriceps aortic
valves, prosthetic valves or with other structural abnormalities like VSD, PDA, and TOF. Aortic root abscess is a life threatening complication of both native and prosthetic valve infections, which requires multidisciplinary management. The cardiologist, intensivist, microbiologist, and surgeon should consult frequently to plan preoperative antibiotic cover, the timing and strategy of surgery, and postoperative management. Despite this, mortality remains high. A patient with recent onset of fever and new heart block should alert the clinician to consider the possibility of infective endocarditis with perivalvular extension of infection or myocardial abscess. The echocardiogram has been used extensively to evaluate patients with endocarditis. The usefulness of all echocardiography techniques is dependent to a large degree on the skill of the person performing the test. Two-dimensional (2-D) transthoracic echocardiography has potential utility in diagnosis of perivalvular abscess.

Our patient’s presentation was unusual because he had a normal aortic valve and he has presented with aortic root abscess with complete heart block and hemodynamic instability. In such cases it is essential to do a screening transthoracic or transesophageal echocardiography to rule out the aortic root vegetation /abscess, so that early surgical intervention can be undertaken to improve chances of survival. New unstable conduction abnormalities in a patient with infective endocarditis mandates aggressive search for perivalvular extension of infection and surgical consultation is advisable in case of persistent conduction abnormalities even after adequate antibiotic therapy for infective endocarditis. Patients with congenital or acquired immunodeficiency syndromes have a greater risk of severe fulminant endocarditis with involvement of multiple cardiac valves. Human immunodeficiency virus may cause aortitis which results in both occlusive and aneurysmal aortic disease. Similarly patients on prolonged steroid therapy, patients with lymphomas and leukemias, those who have been receiving chemotherapy or immunosuppressant therapy are susceptible for severe forms of endocarditis like in this case. All these susceptible individuals need early identification and immediate intervention in the form of broad spectrum antibiotics, supportive therapy and early surgical intervention. Any delay in diagnosis and treatment may prove fatal.

Conclusion
Though common in patients with congenital or rheumatic heart disease, infective endocarditis should be suspected in any patients who present with features consistent with it (tachycardia, bundle branch blocks on ECG and murmur on auscultation). Immunocompromised individuals are highly susceptible to occurrence of endocarditis and hence in these patients possibility of endocarditis and its consequences (valvular involvement, congestive cardiac failure and valvular abscess formation) should be ruled out by appropriate investigations.

Conflict Of Interest: none

Learning Points
Though common in patients with congenital heart diseases, immunocompromised patients and IV drug users, infective endocarditis should be suspected in all patients who present with fever of unknown origin and cardiac conduction defects. Aortic root abscess is a life threatening complication of both native and prosthetic valve infections and requires multidisciplinary management. Early diagnoses of aortic root abscess by transthoracic or transesophageal echocardiography is crucial part of management of these patients. Aggressive medical and surgical management is important as any delay in diagnosis and treatment may prove fatal.

References


