CASE REPORT

Aortic valve replacement in a patient with pancytopenia secondary to myelodysplastic syndrome

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Abstract

The case of a 66-year-old male patient with severe Aortic stenosis who underwent Aortic Valve replacement is presented. This patient was also diagnosed with Myelodysplastic syndrome (MDS) during admission. The perioperative management was challenging because of associated anaemia, pancytopenia and immunosuppression. These patients also have the tendency to bleed not only due to associated thrombocytopenia but also due to the cardiopulmonary bypass related coagulopathy. A thorough preoperative assessment and intensive preparation of the patient was done before taking him to the operating room. There was an effective communication and cooperation between the surgeon, anaesthetist and haematologist at every stage of management which helped in achieving a successful result.

Keywords: Aortic stenosis, Aortic valve replacement, Myelodysplastic syndrome.

Introduction

Myelodysplastic syndrome (MDS) is a clinical condition with progressive pancytopenia, usually caused by dysplasia of the bone marrow. Most patients with Myelodysplastic syndrome develop refractory anaemia and progressive pancytopenia leading to infection and bleeding tendency especially during major surgeries.

These patients may need transfusion of packed red blood cells (PRBCs) and platelets during perioperative period. Use of granulocyte colony-stimulating factor (G-CSF) is also suggested to minimize the risk of infections.[1] Valvular heart surgery itself has a tendency towards coagulopathy and infection and the presence of MDS make them more vulnerable. Extracorporeal circulation (ECC) during cardiac surgery further increases the risk of bleeding.

The perioperative management is challenging in this uncommon disease and preoperative planning is the key to successful management. We present a case of aortic valve replacement (AVR) in a patient with pancytopenia.

Case Report

A 66-year-old, 58kg man was referred for surgical intervention of Aortic valve stenosis. He was suffering from chest pain on exertion and shortness of breath on and off since two years. Physical examination revealed a systolic heart murmur at the 2nd right intercostal space. Electrocardiography showed a sinus rhythm and left ventricular hypertrophy. Echocardiography revealed an aortic valve which was thickened, heavily calcified with severe stenosis, valve area was 0.59 cm/sq, peak pressure gradient of 93 mmHg and severely dilated left atrium. The left ventricular ejection fraction was calculated at 55% with mild to moderate AR. Normal coronary arteries were seen on cardiac catheterization.

Laboratory investigations showed pancytopenia picture with leukocytopenia (white blood cells 3.1×10^9/L), anaemia (haemoglobin 7.7 g/dL) and thrombocytopenia (platelets 127×10^9/L). He was referred to the haematologist for further workup. Peripheral blood film showed dimorphic blood showing anisocytosis and macrocytes, right shifted neutrophils seen and Platelets were low on film. Bone marrow trephine procedure showed blast cells to be less than 5%. Plasma cells were less than 3% and storage iron was missing. Overall cellularity was around 30%. Cellular areas showed the presence of erythroid and myeloid precursors. Dysplastic megakaryocytes were also seen. On the basis of these tests and chromosomal analysis he was diagnosed as Myelodysplastic syndrome (MDS) with refractory anaemia (RA).

His preoperative optimization was performed in collaboration with the haematologist. He was initially treated with injections of Vitamins B12 (four injections) followed by fernous sulphate (Feso4) 200mg BD supplements and folic acid 5mg daily. Normal Chest radiography, liver function tests and viral markers ruled out any chest infection and hepatitis. Preoperative preparation to prevent bleeding and infection included good oro dental hygiene, antiseptic mouthwash, and prophylactic antibiotics.

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On the operation day, Antibiotics, ceftriaxone 1000mg and Vancomycin 1000mg were used prophylactically prior to the skin incision. Tranexamic acid, an antifibrinolytic, 2,500 mg was administered intravenously before the skin incision, followed by 1,000 mg immediately after surgery. AVR surgery was done through a conventional median sternotomy with moderate hypothermia during cardiopulmonary bypass along with antegrade blood cardioplegia. Bypass time was 165 minutes and the aorta cross-clamp time was 120. Heparin was used to maintain ACT of 480 during (CPB) Cardiopulmonary Bypass.

Three units of packed red cell, four units of fresh frozen plasma and six units of platelets were transfused in the operating room. Patient remained haemodynamically stable throughout the procedure. Chest tube drainage was acceptable in the CICU. Fast track extubation was done in the cardiac ICU. Pain was controlled by intravenous tramadol 50mg q6 hourly, Paracetamol I/V 1000 mg q8 hourly and Morphine on PRN basis.

On the day of surgery, total bleeding from his drains was 450 ml at CICU. Chest tubes were removed on 3rd POD and his postoperative course was uneventful. On 5th POD he was discharged home. A peripheral blood examination at discharge still showed pancytopenia, with a white blood cell count of 3.8×10^9/L (neutrophil 69.4%), haemoglobin of 12.4g/dl, and a platelet count 70×10^9/L.

Discussion

Myelodysplastic syndrome (MDS) is a group of clinical disorders which manifests as anaemia, neutropenia and thrombocytopenia of variable severity. It is characterized by ineffective haemopoeisis due to stem cell disorders. Prevalence of this disease increases with age. American and British cooperation group has classified MDS into five categories[2,3] and prognosis can be predicted on the basis of this classification. Patients with Myelodysplastic syndromes are at higher risk of bleeding and infection in major surgeries like coronary artery by pass grafting (CABG), Valvular heart surgery and other major vascular surgeries.

Cardiopulmonary bypass during cardiac surgery is itself associated with changes in coagulation system mainly due to the contact of blood and its component with CPB circuit. These changes include, complement activation, haemolysis, immunological dysfunction, thrombocytopenia and platelet dysfunction. Management of the cardiac surgical patient becomes more complicated and difficult if the patient also has a haemopoeitic disorder like MDS.

In order to avoid perioperative complications, adequate preoperative optimization and planning is vital before proceeding to any elective cardiac surgery in MDS patients. A multidisciplinary team which comprises of a surgeon, anaesthetist, an experienced haematologist and a well-furnished blood bank is recommended for perioperative management of these patients.

MDS is commonly associated with platelet dysfunction[4] and refractory anaemia supplemented by various degrees of granulocytopenia and thrombocytopenia. Although platelet transfusion was not indicated in our patient but due to expected platelet dysfunction associated with MDS and CPB, Platelets were transfused prophylactically to counteract platelet dysfunction. Six units of platelet were transfused after coming off bypass. Antifibrinolytic, tranexamic acid (TXA) has proved to be effective in cardiac surgery patients and this patient also received 3.5 gram of TXA.

Red blood cell transfusion is essential in patients with refractory anaemia to maintain a safe haemoglobin level in patients with cardiac disease undergoing cardiac surgery. Other important measures to help prevent bleeding include careful surgical haemostasis.

Induction, intubation and extubation were smooth and atraumatic in our patient. Strict aseptic precautions were followed for all anaesthetic and surgical maneuvers. Adequate postoperative pain relief was achieved to allow deep breathing exercises, sufficient coughing and hence clearance of respiratory secretions to avoid pulmonary complications and provide smooth and good recovery.

Measures were taken to avoid transmission of infection in the perioperative period. These included aseptic precautions during invasive lines insertion, good oro-dental hygiene when patient was on ventilator, use of new ventilator tubings and use of broad spectrum antibiotics.

These patients also have a higher risk of endocarditis after surgery. This may be the reason that surgeons prefer repair or transcatheter valve interventions (TVI)[5] over replacement.[6] The use of granulocyte colony-stimulating factor (G-CSF) to increase neutrophils count has been mentioned in literature to reduce the number and severity of infections. Previous case reports have mentioned the use of GCSF in cardiac surgery patients[7] but it wasn't suggested by haematologist in our case. Fortunately, he did not develop any infection during his stay in the hospital but prolonged follow up was required.

Conclusion

Present case report shows the classical perioperative

management of Myelodysplastic syndrome for cardiac surgery requiring cardiopulmonary bypass. We were able to manage this patient by required careful planning, coordination among the haematologist, anaesthetist and surgeon and preparation for anticipated complications like infection and coagulopathy.

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