

Clinical Cardiology Updates

Case Report

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Successful Off-Pump Coronary Artery Bypass Surgery in a Patient with Myelodysplastic Syndrome Refractory Anemia with Ring Sideroblasts (MDS RARS): A Rare Case Report [Version 1, 2 Approved]

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Original Submission

Received: November 08, 2016

Accepted: November 16, 2016

Published: November 20, 2016

Last Updated: January 15, 2017

Open Peer Review Status: 2 Approved

How to cite this article: Devender Singh, Shegu Gilbert, Mrinalini Singh. Successful Off-Pump Coronary Artery Bypass Surgery in a Patient with Myelodysplastic Syndrome Refractory Anemia with Ring Sideroblasts (MDS RARS): A Rare Case Report. [Version 1, 2 Approved]. Clin Cardiol Updates. (2016) 1: 1.1

Abstract

Cardiac surgery for patients with myelodysplastic syndrome is very challenging due to the very high risk of complications like bleeding and infection due to the varying degrees of granulocytopenia and thrombocytopenia. Only very few number of patients with myelodysplastic syndrome undergoing cardiac surgery have been reported in literature. Till now only five such patients are reported. We report a 60 year old gentleman with myelodysplastic syndrome refractory anemia with ring sideroblasts who underwent successful off-pump coronary artery bypass surgery. In the post-operative period he developed bleeding due to coagulopathy which was managed appropriately with blood products. He was completely relieved of the symptoms due to coronary ischemia.

Keywords

Myelodysplastic Syndromes; Anemia; Coronary Artery Bypass; Coronary Artery Bypass; Off-Pump

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Introduction

Myelodysplastic syndrome (MDS) refers to a heterogeneous group of closely related clonal hematopoietic disorders usually found in the elderly population. MDS presents with various blood cytopenias. Some of these patients might require coronary artery bypass surgery (CABG) for coronary artery disease during their life time. Patients with MDS have high blood transfusion requirements and run the risk of infections. CABG in these patients is risky due to the complications of anemia, coagulopathy due to platelet dysfunction and sepsis due to neutropenia. Such patients have required multiple transfusions after surgery and some need treatment with granulocyte macrophage colony stimulating factor (GM-CSF). We report the successful off pump CABG in a patient with MDS and the postoperative outcome. Very few patients with myelodysplastic syndrome undergoing cardiac surgery have been reported in the literature. Only five such patients have been reported till now [1-3].

Case Report

A 60 year old gentleman was admitted with history of pain in the left arm and atypical left chest pain since 1 day. He was a diabetic. He also gave history of difficulty in breathing [New York heart association (NYHA) class II] since 6 months. On admission his hemoglobin was 6gms/dL and was diagnosed as non ST elevation myocardial infarction on the basis of positive troponin T test and electrocardiogram (ECG) changes. 2Dimensional echocardiogram (ECHO) revealed anterior wall hypokinesia. He was a known case of myelodysplastic syndrome refractory anemia with ring sideroblasts (MDS RARS) on regular follow up with the hematologist. He had received 6 units of packed red blood cells transfusion in past 2 months and there was no response to erythropoietin injections. His bone marrow trephine biopsy showed hypercellular marrow with megakaryoblastic erythroid hyperplasia and megakaryocytic hyperplasia and more than 20% ringed sideroblasts [Figure 1].

Coronary angiography revealed critical triple vessel disease. As per the American heart association guidelines it is a class IA indication for coronary artery bypass grafting. Since he was anemic he was transfused 2 units of packed red blood cells. His pre-operative platelet count, coagulation, parameters were normal. His pre-operative total leukocyte count was 13500cells/cumm. He was afebrile and there was no evidence of sepsis. He underwent off-pump coronary artery bypass (OP-CAB) with grafts to left anterior descending artery, posterior descending artery, diagonal and obtuse marginal [Figure 2]. Intra operative anticoagulation management was done by giving heparin 2mg/kg prior to coronary artery bypass graft surgery (CABG), maintaining activated clotting time (ACT) of 200 – 250 seconds during grafting and protamine correction was done following completion of the proximal anastomosis. One unit of packed red cells was transfused during the surgery. The total chest drain was 700ml over 24hrs. Hemoglobin on the first post-operative day was 6gm/dL, hence another 2 units of

packed red blood cells were transfused. He required inotropic support (0.1mcg/kg/min) for first 48hrs. He stayed in the intensive care unit for two days. His post-operative course was uneventful and he was discharged home on the 10th day. On the day of discharge his hemoglobin was 8.5gm/dL. He has been on follow up for 3 years and has been stable.

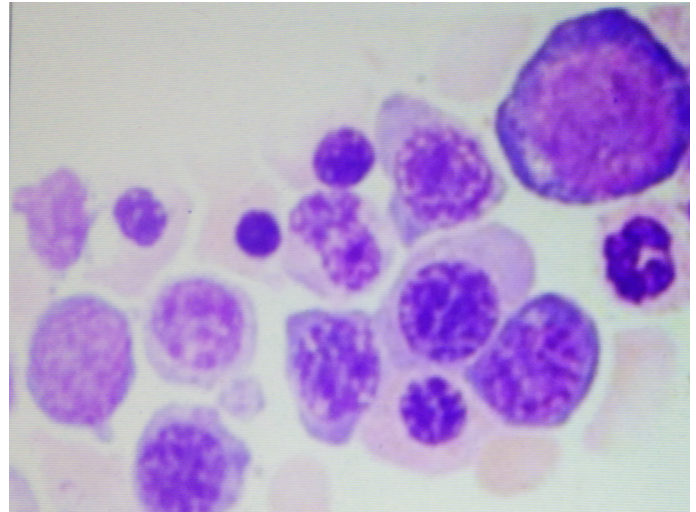


Figure 1: Bone marrow trephine biopsy (H & E stain 100X) shows occasional ring sideroblasts.

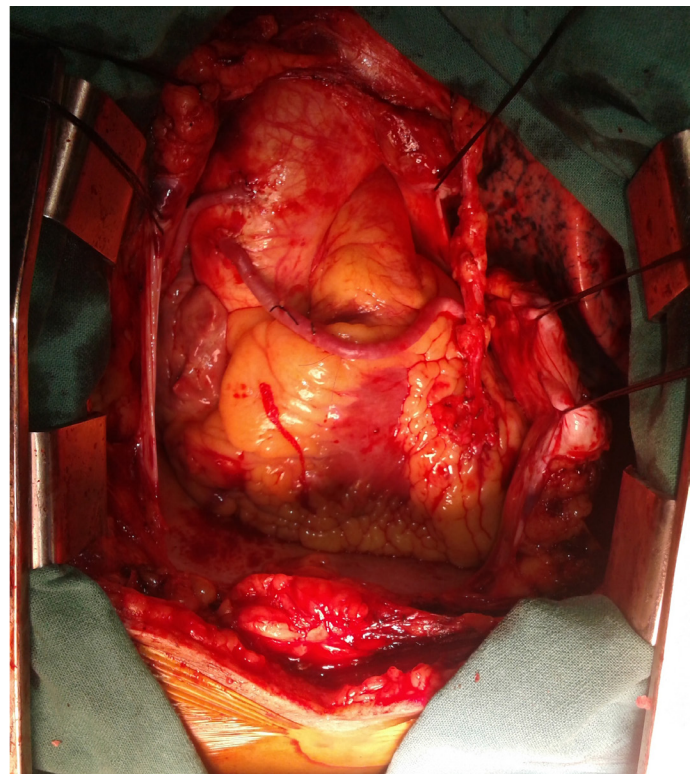


Figure 2: Intra-operative image of CABG surgery.

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Discussion

The myelodysplastic syndrome (MDS) comprise a heterogeneous group of malignant hematopoietic stem cell disorders characterized by dysplastic and ineffective blood cell production and a variable risk of transformation to acute leukemia [4]. Patients with MDS have a variable reduction in the production of normal red blood cells, platelets and mature granulocytes. This often results in a variety of systemic consequences including anemia, bleeding, and an increased risk of infection. WHO classification system distinguishes 6 entities of which refractory anemia with ring sideroblasts accounts for <5% of the cases. RARS is usually associated with good prognosis [5].

CABG in this group of patients is complicated due to anemia, immunological dysfunction and coagulopathy. On pump CABG in this group of patients has been reported earlier. Use of extra corporeal circulation (ECC) in these patients is not without disadvantages. This is due to the exacerbation of the anemia and coagulopathy by ECC. Patients end up requiring massive transfusions which has its own high morbidity. Although Miyagi et al reported lesser need of blood transfusion for their patient who was treated with GM CSF [2]. These patients are prone to hemodynamic instability intraoperatively in view of anemia and may require high inotropic support. Our patient required mild inotropic support for 48hrs. Although the platelet count and coagulation parameters were normal, our patient lost 700ml of blood intraoperatively and another 700ml in the immediate post-operative period. The usual perioperative bleeding in our institution in off pump CABG surgeries is less than 500ml. Patients with MDS also have high incidence of infection related to neutropenia and granulocyte dysfunction [6,7]. In this patient there was no neutropenia and no evidence of sepsis postoperatively. There was no evidence of delayed wound healing. The hemoglobin level during discharge was 8.5gm/dL. Patient has been stable and on regular follow up for 3 years.

Conclusion

MDS is a hematological disorder affecting the elderly population. CABG in these patients is complicated by the risks due to various blood cytopenias. Success of CABG depends on the meticulous preoperative planning, multi-disciplinary team approach, rational use of blood products, use of GM CSF and prevention of infection by use of antibiotics. CABG in these patients can be done with reduced risks. CABG should not be denied for these patients.

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