Anaesthetic Management Of A Case With Sickle Cell Anemia Undergoing Coronary Artery Bypass Grafting

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Abstract

Sickle cell anemia is an autosomal recessive hereditary and hemolytic disorder characterized by abnormal hemoglobin formation (HbS). Surgical interventions carry some risks for sickle cell anemia patients due to the nature of the disease. Our case was a 44-year-old male with sickle cell anemia who underwent coronary artery bypass grafting (CABG) due to coronary artery disease. We report our successful management of anaesthesia and possible pre-, intra-, and postoperative problems for these patients while reviewing the recent literature.

INTRODUCTION

Sickle cell anemia is a hemolytic and hereditary disorder characterized by abnormal hemoglobin formation (HbS). It is transmitted via aoutosomal recessive way. Its acute and chronic symptoms are within the interests of anaesthesiologists ([1,2]). Normal adult red blood cells contain 3 different types of hemoglobin. Hemoglobin A (HbA) consists of 2 and 2 globin chains ($\alpha_2\beta_2$) constituting 96 to 98% of total hemoglobin. Hemoglobin A_2 (HbA_2) consists of 2 and 2 globin chains ($\alpha_2\delta_2$) constituting 1.5 to 3.2% of total hemoglobin. Fetal hemoglobin (HbF), on the other hand, contains 2 and 2 globin chains constituting 0.5 to 0.8% of total hemoglobin. Until the tenth week of life, HbF forms more than 90% of total hemoglobin ([2]). The structure of HbS differs from normal adult hemoglobin (HbA) by substituting glutamic acid at 6th position with valine at 11th position of chain ([1,2]). This mutation is protective against malaria caused by Plasmodium falciparum ([2]). Functionally, affinity of hemoglobin to oxygen and solubility of hemoglobin are less in sickle cell anemia patients. When deoxygenation occurs, HbS polymerizes easily and precipitates within the erythrocytes while making the erythrocytes sickle-shaped ([1,2,3]). Patients form fetal hemoglobin (HbF) in varying amounts (2 to 20%). Cells possessing large amounts of HbF may become protected from sickling to some extent and anemia may become prominent in the 4th month after birth ([1,2]). Continuous formation and destruction of irreversibly sickle-shaped cells cause anemia. Hematocrite level is usually around 18 to 30% due to extravascular hemolysis. In normal human beings, life span of erythrocytes is about 120 days, whereas in patients with sickle cell anemia about 12 to 17 days ([1,3]).

If the genetic disorder of adult hemoglobin originates from both mother and father, the patient is homozygote for HbS and has sickle cell anemia (HbSS). If only one chromosome carries sickle cell gene, the patient is then heterozygote and sickle cell trait (HbAS). Sickle cell trait patients form HbA (55 to 60%) and HbS (35 to 40%) in various amounts. Unlike patients with HbSS, they are usually asymptomatic, not anemic, and have normal life expectancy. Sickling only occurs in cases of when there is excessive hypoxemia or low flow rate. Sickling particularly occurs in renal medulla and most of the sickle cell traits have disturbed renal concentration. It has been reported that some of the patients with HbAS possess renal medullary, splenic and pulmonary infarcts ([1,2,3,4]). Sickle cell anemia essentially is a disease of black race of Middle Africa. About 0,2 to 0,5% of African American people are homozygotic, whereas 8 to 10% are heterozygotic for sickle cell gene. Sickle cell anemia is rarely seen among people of Mediterranean ([1,3]). Situations leading to formation of deoxyhemoglobin such as hypoxemia, acidosis, intracellular hypertonicity or dehydration, increase in 2,3 DPG level or temperature rise may trigger sickling in patients with HbSS. Hypothermia

may cause vasoconstriction and be harmful. Intracellular polymerization of HbS may disrupt the shape of erythtocytes, making them less flexible and more sticky, thus increasing the viscosity of blood. Initially, sickling may be reversible, but may become irreversible in some cells with time. Formation of eryhtrocyte aggregates in capillaries may interrupt with microcirculation of tissues. Circulatory stasis causes localized hypoxia and this increases sickling, leading to a vicious circle. In neonatal period, diagnosis of sickle cell anemia can be made by hemoglobin electrophoresis of umbilical cord blood. Hemoglobin electrophoresis of an infant with sickle cell anemia shows FS pattern (HbF and Hbs). HbF forms 60 to 80% of total hemoglobin. HbA does not exist during neonatal period. In 3rd to 6th months of life, HbF levels fall down to 10 to 20% and HbS is predominant ([3]). Patients with HbSS usually become symptomatic during infancy, where the HbF level decreases obviously. This disorder is characterized by both acute episodic crises and chronic and progressive symptoms. Failure to thrive and recurrent infections are seen in children. Recurrent splenic infarcts lead to splenic atrophy and functional asplenia until adolescence. Patients mostly die of recurrent infections or renal failure. Crises are usually triggered by factors such as infection, cold weather, dehydration or other distressed conditions.

Diagnosis of sickle cell anemia can be made by sickling of erythrocytes following administration of an oxygen consuming substance, metabisulphide or hypertonic ion solution into the medium. Confirmation of this reaction should be made by hemoglobin electrophoresis ([1,2,3,4,5,6]). Moreover, antenatal diagnosis of sickle cell anemia can be made DNA analysis of fetal tissues collected by chorionic villus sampling in first trimester of gestation or by amniocentesis ([2]).

Considering the characteristics of the disorder, surgical procedures may cause unwanted effects among sickle cell anemia cases. Our case underwent coronary artery bypass grafting (CABG) due to coronary artery disease. We discussed our successful management of anaesthesia and possible pre-, intra-, and postoperative problems for these patients while reviewing the recent literature.

CASE REPORT

Our case was a 44-year-old male with sickle cell anemia. He was admitted to Department of Cardiology one month ago with a chief complaint of chest pain. His ECG revealed right bundle branch block and ST elevations in D2, D3 and aVF derivations; and ST depressions in V1 and V2 derivations. He was hospitalized with a diagnosis of acute inferoposterior myocardial infarction. In biochemical investigations results are as follows; blood glucose: 68 mg dL^{-1}, BUN: 12 mg dL^{-1}, creatinine: 1.33 mg dL^{-1}, Na: 138 mEq L^{-1}, K: 4.7 mEq L^{-1}, Ca: 8.1 mEq L^{-1}, Cl: 103 mEq L^{-1}, AST: 113 mg dL^{-1}, ALT: 83 mg dL^{-1}, CK: 578 mg dL^{-1}, CKMB: 183 mg dL^{-1}. His complete blood cell count results are as follows; Hb: 15.9 gr dL^{-1}, Hct: %46.1, WBC: 8800 /mm^3, Plt: 245,000 /mm^3. His coronary angiography revealed 70% stenosis in LAD, 90% stenosis in circumflex artery, 40% stenosis of distal RCA and an ejection fraction of 65%. 3 days later, he was discharged with medications after the relief of chest pain, normalization of biochemical values and hemodynamic stabilization. He was then hospitalized by Department of Cardiovascular Surgery 20 days later for CABG. Results of biochemical tests performed in Department of Cardiovascular Surgery are as follows; glucose: 87 mg dL^{-1}, BUN: 15 mg dL^{-1}, creatinine: 1.11 mg dL^{-1}, Na: 143 mEq L^{-1}, K: 4.3 mEq L^{-1}, Cl: 105 mEq L^{-1}, AST: 17 mg dL^{-1}, ALT: 31 mg dL^{-1}, CK: 16 mg dL^{-1}, CKMB 67 mg dL^{-1}. Results of complete blood count are as follows; Hb: 13.7 gr dL^{-1}, Hct: %40, WBC: 5,500/ mm^3, Plt: 205,000/ mm^3. Hemoglobin electrophoresis showed a HbS count of 35.5%, a HbA count of 50.4%, HbA_2 count of 4% and a HbF count of 1%.

The case was consulted with the Department of Hematology preoperatively. Eryhtropheresis procedure of 5 days duration was carried out. During each sequence, 4 packs of erythrocytes were removed from patient, whereas 4 packs of red blood cell suspension were administered concomitantly.

The operative procedure was chosen as off-pump CABG; as it was the decision of the council. To avoid hypothermia, a warming blanket was put on the operating tablet o warm up the patient. Again, all the administered fluids were heated. For induction of anaesthesia, 100 µg fentanyl, 250 mg sodium thiopental were used and pancuronium was given as muscle relaxant. Sevoflurane within 50% O_2 air mixture and low dose opioid were used for maintenance of anaesthesia. For hemodynamic monitoring, radial artery was cannulated and Swan-Ganz catheter was introduced via internal jugular vein to monitor pulmonary arterial and central venous pressures. Fluid therapy and maintenance of anaesthesia were carried out regarding hemodynamic parameters. Hypoxia and acidosis were avoided via periodic arterial blood gas analyses. There was no need for intraoperative

blood transfusion and our patient was taken to the intensive care unit without experiencing any problem during the surgical procedure. During the postoperative period, fentanyl was used for analgesia and dexmedetomidine for sedation. Arterial blood gas analyses, complete blood count and biochemical tests were within normal limits. He was extubated at 8th postoperative hour. Total drainage through chest tube was 400 milliliters in 24 hours. He was discharged from intensive care unit on 3rd postoperative day.

DISCUSSION

Optimal preoperative preparation should be done for patients with sickle cell anemia. Patients should be hydrated, infections must be controlled and hemoglobin levels should be within acceptable limits. Preoperative need for transfusion should be determined regarding general condition of the patient and surgical procedure he/she will undergo. Partial blood exchange is generally recommended before major surgical interventions in order to minimize sickling by reducing the circulating HbS concentration below 30% ([1,6,7,8]). Blood exchange decreases blood viscosity unlike simple transfusions. Moreover, it increases oxygen carrying capacity and decreases tendency to sickling ([9]). The aim of these transfusions generally is to obtain a hematocrite level of 35 to 40% with a normal hemoglobin (HbA) component of 40 to 50%. Although the benefit of blood exchange in patients receiving anaesthesia is not clearly proven, it certainly is helpful for patients experiencing sickling crises. As mentioned earlier, during preoperative period eryhtropheresis procedure of 5 days duration was carried out. During each sequence, 4 packs of erythrocytes were removed from patient, whereas 4 packs of red blood cell suspension were administered concomitantly. This process provided decreased blood viscosity and increased oxygen carrying capacity.

When reviewing the literature, there were reported cases with sickle cell anemia undergoing on-pump CABG; but we didn't coincide with any case reported undergoing off-pump CABG ([6,7,8,10,11]). In cardiac surgery; cardiopulmonary bypass, aortic cross-clamping, topical hypothermia and cold cardioplegia are all predisposing factors for sickling ([6]). In our case, off-pump method was chosen as more appropriate regarding the coronary lesions and the adverse effects of cardiopulmonary bypass. Conditions causing hemoglobin desaturation and low flow states should be avoided during inraoperative period. All precautions should be taken to prevent hypothermia, hyperthermia, even mild hypoxemia, hypotension and hypovolemia ([1,12]). Adequate hydration and relatively high concentration of inspired oxygen (>50%). The most significant compensatory mechanism for these patients is increase in cardiac output and this should be well preserved during intraoperative period. Monitoring the mixed venous oxygen saturation with central venous and pulmonary arterial pressures may be useful for some patients. Mild alkalosis may prevent sickling but even moderate respiratory alkalosis may have adverse effects on cerebral blood flow. Many clinicians avoid use of tourniquet. There aren't any studies supporting or rejecting any technique of general or regional anaesthesia ([1]). Our case and all the administered fluids were warmed up to prevent hypothermia during the intraoperative period. Fluid therapy and maintenance of anaesthesia were carried out regarding hemodynamic parameters. Hypoxia and acidosis were avoided via periodic arterial blood gas analyses.

The principles that should be taken into account during intraoperative period are also valid for postoperative period. Most of the perioperative deaths ocur during postoperative period. Hypoxemia and pulmonary complications are most significant risk factors. To avoid this type of complications, supportive oxygen therapy, optimal pain relief, pulmonary physiotherapy and early mobilization are necessary ([1]).

Arterial blood gas analyses were repeated during postoperative period, as analgesia and sedation continued. Fast track extubation and oxygen therapy, analgesia and pulmonary physiotherapy after extubation were applied. Our case was discharged from the intensive care unit without any problem on the 3rd postoperative day

In conclusion

Surgical procedures for patients with sickle cell anemia carry risk due to the nature of the disease. Particularly on-pump CABG possesses many complication risks. Especially preferring off-pump CABG method for optimal revascularization in convenient patient groups; multidisciplinary approaches of Departments of Surgery, Anaesthesiology and Hematology; proper anaesthetic management and pre-,intra-, and postoperative precautions could prevent these complications.

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