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Case Report

Sickle Cell Trait and Mitral Valve Surgery: A Report of Two Cases

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Abstract

Patients with sickle cell hemoglobinopathies who require cardiac surgery are at risk of a potentially fatal sickling crisis, which may be triggered by factors such as hypothermia, hypoxia, acidosis, or low-flow states. Clinically, this is manifested as episodic vascular occlusion leading to tissue ischemia and end-organ damage. Consequently, major surgeries involving cardiopulmonary bypass and hypothermia are particularly risky for these patients. This report discusses the management of two patients with sickle cell trait who successfully underwent mitral valve replacement.

Kew Words: sickle cell trait, mitral valve replacement, cardiopulmonary bypass

Introduction

Sickle-cell disease (SCD) affects more than 30 million people globally, with a higher incidence among populations of African descent, including Afro-Caribbeans, as well as in the Mediterranean Basin, the Middle East, and India. [1] It results from a mutation at the sixth position of the β-globin gene, which causes substitution of glutamic acid with valine. The autosomal recessive mutation of the β-globin gene produces an abnormal hemoglobin (Hb), called 'S' (HbS).[2] Low oxygen tension causes HbS polymerization, which is responsible for erythrocytes' deformation into an irregular sickle shape. Stiffness in sickle cell walls leads to microvascular occlusion, reperfusion injury, infarction, chronic hemolysis, endothelial dysfunction, and inflammatory vasculopathy, causing a multi-systemic involvement.[1] Sickle cell hemoglobinopathies can range from the usually benign sickle cell trait [SCT] to the potentially fatal sickle cell anemia.

Patients with SCD who require cardiac surgery are at risk of a potentially fatal sickling crisis, which may be precipitated by hypothermia, hypoxia, acidosis, low-flow states, dehydration, stress, inflammation, or infection. [3] Clinically, this is manifested as episodic vascular occlusion leading to tissue ischemia and end-organ damage, making major surgery involving cardiopulmonary bypass (CPB) and prolonged anesthesia a greater risk in this population. We report two cases of mitral valve replacement (MVR) in patients with SCT.

Case Description

Two African women, aged 51 and 58 years, with a history of rheumatic fever and recurrent heart failure requiring hospitalization, developed progressive dyspnea and fatigue (New York Heart Association class III). Echocardiogram revealed severe mitral stenosis with a mean gradient of 18 mmHg in patient 1 and an eccentric jet of severe mitral regurgitation in patient 2. The sickling test was positive in both patients. Hemoglobin electrophoresis was performed by high-performance liquid chromatography (HPLC) to detect the concentrations of HbS, HbC, and HbA. It showed HbS values of 24.5% (patient 1) and 36.8 % (patient 2). Mitral valve replacement was recommended for both patients. The details of patients' demographics, laboratory results, and surgical procedures are provided in Table 1. During a six hours period of nil per os, 5% dextrose in saline was administered, and oral alprazolam 0.5 mg was administered 45 minutes before the scheduled procedure. A balanced anesthetic technique consisting of midazolam, fentanyl, thiopentone, vecuronium, and isoflurane was used for surgery. Intraoperative monitoring was standard and consisted of invasive arterial pressure, transesophageal echocardiography, pulmonary artery catheterization, and cerebral oximetry. Cerebral oximetry values were kept above 55%. After aortic and bicaval cannulation, but before institution of cardiopulmonary bypass (CPB), partial exchange transfusion was performed by draining 500 ml of the patient's blood and by replenishing it with two units of healthy red blood cells and plasma obtained from the blood bank. Blood from the cardiotomy suckers was discarded, the cell saver was not used, and auto-transfusion was not performed. During CPB, core temperature was not allowed to drift below 35^0C. After application of the aortic cross clamp, one shot of warm blood cardioplegia was injected to flush out the existing blood in the coronaries and reduce sickling in the coronaries. This was followed by a full dose of cardioplegia (20 ml/kg), and a repeat dose of 10 ml/kg every 20 minutes. Intraoperative hypothermia was avoided by the Bair Hugger™ warming device, warm fluids, warming blankets, and ambient theater temperature. Propofol and fentanyl infusions were used for postoperative sedation and analgesia. The patients were extubated on postoperative day 1, and the remaining course after surgery was unremarkable.

Test	Patient 1	Patient 2

Age (years)/Gender	51 / Female	58 yrs / Female
Height/Weight/BSA (cm)/(kg/(m^2))	160/51/1.51	170/81/1.93
Hb (gm/dl)/hematocrit (%)	9.8/29.0	8.3/26.4
Hb Electrophoresis (HPLC)	HbA 61.3% HbA_2 4.3% HbS 24.5% HbC 1.1% HbF <0.8% HbA_{1c} 6% P_3 4%	HbA 48.5% HbA_2 3.8% HbS 36.8% HbC 0% HbF <0.8% HbA_{1c} 6.1% P_3 4%
Diagnosis	Severe MS, moderate TR, Pulmonary hypertension	Severe MR, Coronary artery disease
Co-morbid conditions	Pulmonary tuberculosis	AKI (S. Creatinine 2.2 mg/dl)
EuroSCORE II	1.68%	6.26%
Aortic cross clamp time	79 min	67 min
CPB time	96 min	95 min
Surgery	MVR (25 mm EPIC bioprosthetic valve)	MVR (25 mm SJM mechanical valve) + CABG x 1
Postoperative transvalvular pressure gradient	2.3/ 5.6 mmHg (mean/peak)	1.4/ 4.8 mmHg (mean/peak)
RBC/FFP/Platelets	4/2/--	7/6/4
Hemolytic crisis	No	No
ICU readmission	No	Yes (for pericardial and pleural tapping)
Hospital stay	11 days	18 days

Table 1: Patients' demographics, laboratory results, and surgical procedures

Abbreviations (Table 1): BSA-body surface area, Hb-hemoglobin, HPLC- High-performance liquid chromatography, MS-mitral stenosis, TR-tricuspid regurgitation, MR-mitral regurgitation, AKI-acute kidney injury, CPB-cardiopulmonary bypass, MVR-mitral valve replacement, CABG-coronary artery bypass grafting, RBC-red blood cells, FFP-fresh frozen plasma, ICU-intensive care unit

Discussion

Sickle cell disease is a hereditary hemoglobinopathy resulting from the inheritance of a mutant version of the globin gene on chromosome 11. The condition may present as SCD, the severe form of which is the homozygous genotype (HbSS), in which the fractional concentration of HbS ranges between 70% and 98%, or it can be manifested as SCT, which is rather benign and more common among populations as the heterozygous genotype (HbAS), in which the fractional concentration of HbS is <50%. [4] CPB has been performed on SCD and SCT patients using various aggressive transfusion strategies before or during bypass, designed to decrease the proportion of HbS.[5] There is a lack of definitive control data to validate the need for these techniques. In more recent series, a total of 28 SCT patients tolerated bypass without alterations to standard protocol. [6] Transfusion was avoided where possible. Preventative transfusion, therefore, does not appear to be necessary for SCT patients. There are case reports of successful hypothermic bypass without preoperative transfusion in four patients with SCD (two with a blood prime and two who did not receive any transfusion before or during bypass).[7] Bypass without transfusion is feasible in SCD, although there are inadequate data to accurately quantify the risks. For patients at low risk, any potential benefit derived from transfusion would be minimal and offset by a high incidence of complications. [5] Erythrocyte transfusion is therefore not indicated. For patients at moderate or high risk, the potential benefit of transfusion might be expected to be greater. If undertaken in situations of moderate risk, transfusion should aim for a hematocrit of 30% rather than aim to achieve a target dilution of HbS. The timing of prophylactic transfusion can vary from the immediate preoperative period to up to a week before scheduled surgery. Exchange transfusion can be done preoperatively, perioperatively, or both. Although there is no consensus on the absolute safe values of HbS in patients undergoing surgery, it is proposed that the level of HbS should be reduced to 30% for major surgical procedures. [4] Exchange transfusion is particularly relevant in the context of deep hypothermic circulatory arrest, when the risk of sickling is so much higher. Priming the bypass circuit with sickle-negative blood allows the patient's sickled blood to be simultaneously replaced before CPB is established, and hence the procedure can usually be undertaken without inducing haemodynamic instability. Such a strategy was employed by many authors, including Taylor et al, in acute type A aortic dissection, which required core cooling to a temperature of 18^0C. [9] The patient did not develop sickle cell crisis or any other major complications.

CPB has been performed in patients with all hemoglobin phenotypes. Abnormal cardiac physiology is usually secondary to chronic anemia, advanced pulmonary pathology, or coexistent structural cardiac abnormalities, rather than a specific SCD-induced cardiomyopathy. Large vessel coronary arteriosclerosis is not a complication of SCD. Indications for CPB include valve and congenital defect repairs, pulmonary thrombectomy, and coronary artery bypass grafting. Prosthetic valves were not associated with excessive hemolysis in SCD or SCT patients. Systemic hypothermia, aortic cross clamping, acidosis, low flow states, and cold cardioplegia during CPB have variously been suggested as potential precipitants of sickling crisis. [10] It should be noted that the above-mentioned predisposing conditions are more common in patients undergoing cardiac surgery, CPB itself, as well as topical or whole-body hypothermia, cold cardioplegia, and use of vasoconstrictive agents, may predispose to the crisis state. Hence, special care should be taken in sickle cell patients who require cardiac surgery to avoid or at least to minimize those risk factors. The potentially catastrophic consequences of sickling in patients undergoing cardiac transplantation are also reported.[10] SCT is not a benign condition at the extremes of physiology encountered during CPB and requires careful assessment and consideration of the strategies required to minimize the risk of a perioperative sickle crisis. However, bypass takes place under conditions of deep anesthesia, hemodilution, and profound anticoagulation designed to minimize these physiologic insults. Conceptually, complications of SCD would probably be triggered by the large post-bypass inflammatory response rather than by the mechanics of erythrocyte sickling in the bypass circuit. Sickling may occur in the heterozygous state at a PaO_2 of 20-25 mmHg, whereas

in the homozygous form it occurs at a higher PaO_2 of 40 mmHg, emphasizing the importance of avoiding hypoxia. To decrease the risk of sickling, the reduction of HbS% by partial exchange transfusion immediately before CPB has been suggested. This technique was used in the cases described here. A recent study of 20 patients with SCD and 40 patients without SCD, has shown similar non-SCD-related complications in both groups and no mortality in either group.[8] This might necessitate the use of blood in the priming solution, which alone would be sufficient to decrease the percentage of HbS. Basic supportive care, including adequate analgesia, incentive spirometry, early mobilization, and oxygen supplementation as needed to prevent hypoxemia, is the mainstay of postoperative management. [5]

In conclusion, cardiac surgery using CPB can be performed successfully in patients with sickle cell hemoglobinopathies. Potentially fatal sickling crisis can be prevented by avoiding the precipitating factors such as hypothermia, hypoxia, acidosis, low-flow states, dehydration, and stress

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