Reoperative Hemoglobinuria in a Coronary Artery Bypass Graft Case with Hereditary Spherocytosis

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Abstract

We present a rare case of hereditary spherocytosis (HS) with intraoperative hemolysis. A 60-year-old man with coronary artery disease, HS, and a history of splenectomy for HS and underwent coronary artery bypass graft surgery under cardiopulmonary bypass, during which he developed severe hematuria and hemolysis and his hemoglobin reached 5g/dL***(Iranian Heart Journal 2011; 12 (1):63-66).***

Keywords: hereditary spherocytosis■ hematuria■ coronary artery bypass graft surgery

Open-heart surgery for acquired cardiac lesions in patients with hematologic diseases such as inherited hemoglobinopathies and red cell dyscrasias presents potential management problems during the perioperative or postoperative period.

Among these disorders, hereditary spherocytosis (HS) is an intrinsic red blood cell defect which results in hemolytic anemia with a prevalence rate between 0.02% and 0.05% of the population in Europe and North America.[1] In HS, the red blood cells are spheroidal in shape and have an increased osmotic and mechanical fragility. Meanwhile, the use of cardiopulmonary bypass (CPB) pump during a cardiac operation causes some inevitable hemolysis, platelet destruction, and protein denaturation, which sometimes leads to fatal hematologic conditions.

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We herein report the case of a 60-year-old man with HS, who developed severe intraoperative hemolysis during coronary artery bypass graft surgery (CABG).

Case Report

A 60-year-old man (70kg in weight and 165cm in height) with a history of chest pain for the previous four months was referred to our preoperative evaluation clinic, where coronary artery disease was diagnosed and CABG was scheduled. The patient had a history of splenectomy forty-five years previously for HS. His father also had HS. Before surgery, pneumococcal vaccine was administered. Preoperative laboratory test results were as follows:

Hb:13.9 g/dl, HCT: 39.9%, MCV: 75fl, MCH: 26Pg, MCHC: 35g/dl, Bill=1mg/dL,

Direct 0.3, indirect:0.7, and PLt: 479000/cumm. Other laboratory tests were normal. The patient consumed carvidilol 6.25mg Bd., Lozar 12.5mg Bd., and atorvastatin 40mg daily.

There was no significant finding on carotid artery Doppler sonography. Echocardiography revealed left ventricular (LV) enlargement, LV systolic and diastolic

The patient continued his cardiac medication on the morning of surgery. Premedication comprised oral lorazepam (1mg), administered during the night before the operation and one hour before anesthesia induction, and intramuscular morphine (5mg), also administered one hour before induction.

In the operating room, the patient was monitored via standard ECG (V5, II), pulse oxymetry, and nasopharyngeal core temperature. The BIS (SpaceLabs Medical Model No 90491) and then the veins on both arms were cannulated with 16-g catheters, under local anesthesia with 1% lidocaine. Left radial artery cannulation was done (20-G catheter arrow REF SAC 00820), and the central venous catheter (Arrow-Howestm multi-lumen central venous catheterization set

with Blue flex Tip® catheter No 7Fr) was placed into the right subclavian vein. After recording the baseline measurements, anesthesia was induced using fentanyl 10µg/kg, etomidate 0.2mg/kg, and cisatracurium 0.2 mg/kg. The patient was ventilated manually with 100% oxygen until intubation was done. Thereafter with intermittent positive pressure ventilation (MEDEC, Saturn EVO), the end-tidal Co2 partial pressure was maintained at 35-40mmHg. Anesthesia was maintained with oxygen 100%, propofol 50µg/kg/min, fentanyl 1-2µg/kg/h, and atracurium 0.6mg/kg/h. The doses of the intravenous anesthetic agents were primarily adapted to hemodynamic tolerance. The BIS values were maintained between 40 and 60.

The patient had a median sternotomy. A standard cannulation technique was employed to perform CPB, in which cannulae were placed in the ascending aorta and right atrium. CPB was initiated after systemic heparinization (300IU/kg); the activated clotting time (ACT) was more than 480 seconds. Extracorporeal circulation was performed with a standard membrane oxygenator (Medtronic. TRILLLIUM®. AFFINITY® NT). The pump flow rate was maintained between 1.5-2 L/m^2. During the extracorporeal circulation, the mean arterial pressure was maintained between 40 and 60 mmHg. Myocardial protection was provided through mild systemic hypothermia (32^{oc}), topical ice, and intermittent antegrade cold cardioplegia delivered via a cannula placed in the aortic root.

After thirty minutes of CPB, the patient developed hematuria and his Hb reached 5gr/dl. Furosemide 60mg and bicarbonate 70mEq were administered and also 250mL packed cell and 200mL manitol (20%) were added to the prime.

Meanwhile, the surgeon constructed anastomosis; and the patient with an intra-aortic balloon pump (DataScope CS10 Intelligent Counterpulsation) 1:1 and adrenaline 0.01 µg/kg/min was separated from CPB at the second attempt. The total CPB time and clamp time were 135 minutes and 48 minutes, respectively. During CPB, ultrafiltration was done and the urine output was measured at 1200mL. After surgery, the patient was transferred to the intensive care unit (ICU) while receiving epinepherin 0.01µg/kg/min and IABB 1:1. The patient's mean arterial pressure was 60-70mmHg. Serial ABGs were conducted, and bicarbonate and furosemide was administered in order to balance the urine PH between7.5- 8 and maintain diuresis on at least 75-100mL/hour. Additionally, 5 units of packed red cells were administered to increase Hb to 10mg/dL. During the first postoperative day, eight units of packed cells were administered in order to maintain the Hb level between10-11 gr/dL. On the second day of admission to the ICU, epinephrine was tapered, IABP decreased to 1:2 and then 1:3, urine color became clear, urine hemoglobin decreased, and the serum hemoglobin level remained steady. Forty hours after the operation, the patient was extubated. No hemodynamical and renal complications were detected three days after extubation, and the patient was finally discharged on the eighth postoperative day. At six months' follow-up, there was no evidence of renal, hematologic, cerebral, or cardiac complications and the laboratory test results were as follows:

Acanthocytosis, mild anisopoikilocytosis, mild spherocytes, Howelljolly bodies, and Hb: 12.1g/dl.

Table I. Perioperative and postoperative plasma hemoglobin concentrations

Parameters	Measured plasma hemoglobin g/dL
Preoperation	13.9
Just before CPB	13
After the beginning of CPB and hematuria	5

At the termination of CPB	8.5
Just admitted to ICU	7
24 hours after admission to ICU	10

CPB: cardiopulmonary bypass

Discussion

HS is most often transmitted by autosomal-dominant inheritance, but recessive and isolated mutation forms of transmission also have been described.[2] The disease is characterized by the presence of spherical–shaped erythrocytes in the peripheral blood smear, reticulocytosis, and enhanced red blood cell fragility concomitant with elevated serum bilirubin concentrations. [3]

Spherocytes lack the deformability and elasticity of normal erythrocytes because of the presence of defective membrane structural proteins and, as a result, the life span of these abnormal red blood cell is substantially reduced by mechanical and shear stress, hemolysis, and subsequent splenic sequestration and dectruction.[4] On the other hand, some degree of hemolysis is unavoidable in open-heart surgery under CPB. This is one of the major concerns in open-heart surgery. Patients with HS have an accentuation perioperative risk of hemolysis caused by fragility of erythrocytes.[5] Very mild forms of HS may be entirely asymptomatic. Patients with severe disease often display profound anemia (hemoglobin concentration < 8g/dL) and thus require frequent blood transfusion. In addition, they tend to develop splenomegaly, unconjugated indirect hyperbilirubinemia, jaundice, and cholelithiasis as a result of excessive hepatic bilirubin metabolism; they may also suffer hemolytic, megaloblastic, or aplastic crisis in conjunction with viral infection.[6-7]

These clinical manifestations of HS are substantially mitigated by splenectomy in the majority of patients; splenectomy is recommended before cardiac operations to prevent significant hemolysis.[7-8] Initial descriptions of cardiac surgery in patients with HS appeared in the early 1970s; and to date, no perioperative mortality has been attributed directly to this hemolytic anemia.[9] Moyes et al. first reported mitral valve and Tetralogy of Fallot repairs using CPB in 2 children with HS.[10] Excessive perioperative hemolysis occurred in a 67-year-old man with HS who underwent an aortic valve replacement; the patient eventually required splenectomy for the treatment of severe hemolytic anemia attributed at least in part to the implanted mechanical prosthesis and not to the exposure to CPB per se.[11] It is also recommended that mechanical valve prostheses be relatively contraindicated in these patients based on experience.[11] An off-pump approach to direct myocardial revascularization that avoids CPB entirely may be beneficial in patients with severe HS-induced hemolytic anemia and coronary artery disease.

In our case, severe hemolysis occurred due to contact between defective red blood cell membrane and the oxygenator membrane. The objective for this patient was an intensive care plan so as to prevent renal failure and to maintain Hb serum at a minimum level of 10gr/dl; this was achieved through a proper diuresis. The alkalinization of the urine to prevent the precipitation of acid hematin in the distal tubules is of questionable value but is easy and therefore recommended in tandem with packed red blood cell administration.

The risk of both arterial and venous thromboembolism is elevated long term after splenectomy in this patient population,[12] be that as it may, our findings were similar to those reported in asplenic patients who had undergone splenectomy for other indications.

It should be noted that these complications did not occur in our patient.
Although mortality as a result of HS in the wake of cardiac surgery is very rare, hemolysis could occur.

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