DOI: 10.7860/JCDR/2016/23379.9073

Case Report

Surgery Section

Precautionary Measures for Successful Open Heart Surgery in G6PD Deficient Patient- A Case Report

RUPESH KUMAR

ABSTRACT

Glucose-6-Phosphate Dehydrogenase (G6PD) deficiency is among the most common enzymatic disorders of red blood cells. Cardiac surgeries on this group of individuals are associated with an additional risk in terms of impaired oxygenation, prolonged ventilation and increased risk of haemolysis. These patients have a very low threshold for haemolysis due to oxidative stress. Many commonly used drugs also predispose the individual for haemolysis when they are subjected to surgery. Here we present a known case of G6PD deficient patient with symptoms of breathlessness for the last nine years who was taken for surgery with pre-planned precautionary measures to avoid unnecessary haemolysis. The echocardiography report revealed severe mixed mitral lesion and moderate tricuspid regurgitation. On general examination she had mild pallor and icterus. We planned for a thorough investigation to prepare her for mitral valve replacement and tricuspid annuloplasty. These groups of patients are at high risk of haemolysis during perioperative period and need prolonged mechanical ventilation and hospital stay due to impaired oxygen carrying capacity and oxidative stress due to deficient free radical scavenging system. The patient underwent mechanical mitral valve replacement and tricuspid annuloplasty under cardiopulmonary bypass with precautionary measures to prevent the risk of haemolysis and associated complications. She had an uneventful recovery.

Keywords: Glucose 6 phosphate dehydrogenase, Haemolysis, Red blood cells

CASE REPORT

A 30-year-old lady attended our hospital with chief complaints of symptoms of breathlessness and yellowish discolouration of eyes for the last nine years. She was a known case of G6PD deficiency. On general examination, she was afebrile, icteric with no organomegaly or pedal oedema. Routine blood tests revealed high total bilirubin level (5.01 mg/dl) with predominantly indirect hyperbilirubinemia, the aspartate aminotransferase level was 47.72 IU/L. Abdominal ultrasonography was grossly normal. We reassessed the qualitative as well as quantitative analysis of G6PD and found her to be severely deficient with blood level of G6PD as 0.10 U/gHb (normal range-4.6-13.5 U/gHb).

Echocardiography report suggested rheumatic heart disease involving mitral valve causing severe mitral stenosis and regurgitation and moderate tricuspid regurgitation and pulmonary arterial hypertension with left atrial enlargement and a left ventricular ejection fraction of 45%.

The patient was prepared for surgery and she received tablet furosemide 40mg, a day before surgery to ensure good urine outflow to avoid a renal tubular injury due to the free haemoglobin. Premedication was instituted with intravascular glycopyrrolate 0.2 mg followed by induction with propofol 150mg and fentanyl 150 µg. Endotracheal intubation was facilitated with rocuronium 60 mg. Anaesthesia was maintained with isoflurane, O_2/N_2O mixture, rocuronium and fentanyl.

A median sternotomy was performed followed by pericardiotomy and external anatomy of heart was assessed. Following systemic heparinisation, Cardiopulmonary Bypass (CPB) was instituted with aortic and bicaval cannulation and antegrade cardioplegia was administered every 15 minutes for myocardial preservation. We used roller pump with minimum occlusion to avoid haemolysis. Albumin was used in prime as coating of CPB circuit with albumin prevents initiation of complement cascade. Through a left atriotomy approach, chordal preserving mitral valve replacement with 27 mm bileaflet mechanical mitral heart valve prosthesis using 2-0 pledgeted polyester sutures was done. De Vega annuloplasty was performed for tricuspid regurgitation. The procedure was conducted under normothermic bypass while maintaining a mean blood pressure of 65 mm of Hg. The procedure happened uneventfully and the patient was weaned off CPB. The aortic cross clamp time was 75 minutes and the CPB time was 100 minutes.

No extra blood products were transfused to the patient and the colour of the urine was amber without any signs of haematuria. The patient was shifted to the ICU and was extubated within six hours of shift to the ICU. The pericardial drain output was 90 ml in 24 hours.

The patient had no episode of haematuria during the hospital stay and was discharged on the fifth postoperative day. She is on regular follow-up for the last six months without any ailments.

DISCUSSION

G6PD deficiency is one of the most common enzymatic disorders of red blood cells with a prevalence of about 400 million cases worldwide [1]. G6PD is an enzyme that catalyses the first step in hexose monophosphate pathway of glucose metabolism which leads to production of reduced Nicotinamide Adenine Dinucleotide Phosphate (NADPH), which helps in maintaining reduced glutathione necessary for protecting the red blood cells from oxidative damage [2]. CPB procedures cause inflammatory reactions, oxidative stress as well as haemolytic derangements. So, such surgeries are considered to have a high risk for perioperative morbidity [3]. G6PD is an enzyme in the hexose monophosphate pathway and its deficiency is responsible for multiple consequences in human body. The deficiency of this enzyme leads to signs and symptoms like anaemia, jaundice, hepatosplenomegaly, reticulocytosis and all consequences of corpuscular haemolysis [4]. Drugs that lead to peroxide release cause oxidation of haemoglobin and red blood cell membranes and aggravates the symptoms of G6PD deficiency. The diagnosis of G6PD deficiency is made by spectrophotometry which detects the generation of NADPH from NADP [5]. These patients are susceptible to oxidative stress manifesting as haemolysis when exposed to infections, certain precipitating drugs and

extracorporeal circuits [6]. During CPB the oxidative stress along with the mechanical trauma from the roller pumps and cardiotomy suction leads to haemolysis, endothelial cell injury and capillary leaks during the post bypass surgery period [7]. Vital organs like lungs and kidneys are at most risk of injury [8]. In cardiac surgery these risks are compounded by more commonly used drugs like paracetamol, aspirin, sulphonamides and nitrates [9].

CONCLUSION

A careful workup, in perioperative period and strategies to minimise haemolysis by quick surgery, less CPB and aortic cross clamp time, maintenance of normothermic bypass, avoidance of oxidative drugs and maintaining good urine output leads to successful perioperative management of such types of cases.

REFERENCES

[1] Glader BE. Glucose-6-phosphate dehydrogenase deficiency and related disorders of hexose monophosphate shunt and glutathione metabolism. In: Wintrobe's Clinical Haematology. 10th ed. Baltimore: Williams & Wilkins; 2008: 1176–90.
[2] Luzzatto L, Nannelli C, Notaro R. Glucose-6-phosphate dehydrogenase deficiency. *Haematol Oncol Clin North Am*. 2016;30(2):373-93.
[3] Gerrah R, Shargal Y, Elami A. Impaired oxygenation and increased haemolysis after cardiopulmonary bypass in patients with glucose-6-phosphate dehydrogenase deficiency. *Ann Thorac Surg*. 2003;76:523–27.
[4] Tas S, Donmez AA, Kirali K, Alp MH, Yakut C. Aortic valve replacement in patient with glucose-6-phosphate dehydrogenase deficiency and autoimmune haemolytic anemia. *J Card Surg*. 2005;20:380-81.
[5] Kaplan M, Hammerman C. Glucose-6-phosphate dehydrogenase deficiency and severe neonatal hyperbilirubinemia: a complexity of interactions between genes and environment. *Seminars in Fetal & Neonatal Medicine*. 2010;15:148–56.
[6] Maddali MM, Fahr J. Postoperative methaemoglobinemia with associated G-6-P-D deficiency in infant cardiac surgery – enigmas in diagnosis and management. *Pediatric Anesthesia*. 2005;15:334–37.
[7] Chowdhry V, Bisoyi S, Mishra B. Perioperative challenges in a patient of severe G6PD deficiency undergoing open heart surgery. *Ann Card Anaesth.* 2012;15:50-53.
[8] Elyassy AR, Rowshan HH. Perioperative management of Glucose-6-Phosphate Dehydrogenase deficient patient: A review of literature. *Anesth Prog*. 2009;56:86-91.
[9] Younster I, Arcavi L, Schechmaster R, Akayzen Y, Poplisky H, Shimonov J. Medications and glucose-6-phosphate dehydrogenase deficiency: An evidence based review. *Drug Saf*. 2010;33:713-26.

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Cardiothoracic and Vascular Surgery, Advanced Cardiac Center, Post Graduate Institute of Medical Education and Research, Sector 12, Chandigarh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Rupesh Kumar,
Assistant Professor, Department Of Cardio-Thoracic And Vascular Surgery, Advanced Cardiac Center,
Room 4018, 4th Floor, PGIMER, Chandigarh-160012, India.
E-mail: rkctvs@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 31, 2016**
Date of Peer Review: **Sep 16, 2016**
Date of Acceptance: **Sep 26, 2016**
Date of Publishing: **Dec 01, 2016**