Case report

Successful CABG in a patient with paroxysmal nocturnal hemoglobinuria

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Abstract

Paroxysmal nocturnal hemoglobinuria (PNH) is an uncommon acquired disorder of blood cells characterized by intravascular hemolysis due to an inordinate sensitivity to complement-mediated membrane damage. This report documents coronary artery revascularization in a patient with PNH, which has not been described in the literature so far. We used the on-pump procedure in the present patient and successfully performed revascularization by taking prophylactic measures against hemolysis.

Keywords: Paroxysmal nocturnal hemoglobinuria; Coronary artery bypass grafting; Complement; Hemolysis; On-pump

1. Introduction

As cardiac surgery has evolved, more complicated cases are being seen. Paroxysmal nocturnal hemoglobinuria (PNH) is an uncommon acquired disorder of blood cells related to the proliferation of abnormal clonal stem cell population. It is principally characterized by intravascular hemolysis due to an inordinate sensitivity to complement-mediated membrane damage. This report documents coronary artery revascularization in a patient with PNH, which has not been described previously in the literature.

2. Case report

The patient was a 78-year-old man who had been referred to our hospital for coronary artery revascularization. A diagnosis of PNH had been made 3 years before the hospitalization for cardiac surgery and later was confirmed by flow cytometry (Fig. 1). He seemed to have suffered from hemolysis for 3 years and medical therapy included corticosteroid therapy (20 mg/day) that was complicated by retrogression of diabetes mellitus. On admission, laboratory tests exhibited normal kidney function (blood urea nitrogen of 15 mg/dl and creatinin of 0.7 mg/dl) and normal liver function (glutamic pyruvic transaminase of 14 IU/l, cholinesterase of 85 IU/l, and platelet count of 44.3 x 10^5/µl). Hemolytic attack occurred 5 days before surgery and laboratory studies showed a normocytic and normochromic anemia with a hemoglobin value of 8.0 g/dl, and enzyme analysis revealed elevated glutamic oxaloacetic transaminase of 51 IU/l and lactate dehydrogenase of 1352 IU/l. He received blood transfusion of concentrated red cells and his anemia was ameliorated temporarily. His chest X-ray showed moderate cardiomegaly (cardiothoracic ratio of 0.67), and his electrocardiogram showed Q waves in leads II, III, and aVF, consistent with an old inferior wall infarction. Coronary angiography revealed a total occlusion of the left anterior descending (LAD) coronary artery with retrograde filling from the right coronary artery (RCA). There were 90% stenoses in the proximal segment of the left circumflex coronary artery (LCX) and in the middle portion of the RCA. Left ventricular angiography showed severe hypokinesis of the posterior wall with an ejection fraction of 35%.

General anesthesia was induced with fentanyl and midazolam, and the patient was paralyzed with vecuronium. Anesthesia was maintained with oxygen, and incremental doses of fentanyl and midazolam. Coronary artery revascularization was performed with the aid of cardiopulmonary bypass (CPB). A left internal mammary artery and saphenous vein graft were anastomosed to the LAD and the posterior branch of the LCX, respectively, under cardioplegic arrest, and the proximal anastomosis of
the saphenous vein graft was constructed under partial clamping of the aorta.

The patient was weaned from CPB without difficulty. Slight intraoperative hemoglobinuria was noted but disappeared by the end of the operation.

To prevent hemolytic crisis, we performed the following prophylactic management during the perioperative period:

1. Preoperative transfusion of group-specific red cells with white cell depletion by filtration in order to decrease abnormal red blood cells.
2. Infusion of corticosteroid intravenously at a dose of 100 mg before induction of anesthesia, 1 g on the initiation of CPB, and 50, 50, and 20 mg on the first, second, and third postoperative days, respectively.
3. Use of haptoglobin (4000 U) during CPB for prevention of renal failure in case acute hemolysis occurred.
4. Administration of low molecular weight dextran in the intensive care unit.

The postoperative course was uneventful, and hemoglobinuria did not occur during hospitalization (Fig. 2).

3. Discussion

Hemolysis associated with PNH occurs because of an abnormality of the red cell membrane that makes the cells unusually susceptible to the lytic action of complement. When complement is activated via either the classical or the alternative pathway, it results in a continuous deposition of complement components, especially C3b, onto the erythrocyte surface causing hemolysis [1].

The main conundrum in this case was to decide which method, on-pump coronary artery bypass grafting (CABG) or off-pump CABG, should be used to avoid the hemolysis associated with PNH. Many studies have shown that off-pump coronary artery bypass surgery results in lower activation of inflammatory mediators than does conventional on-pump coronary artery bypass surgery [2]. However, Fransen and associates [3] indicated that the acute phase response in CABG patients, which has historically been ascribed to the CPB procedure, is predominantly caused by the surgical procedure per se. Recent data from Tarnok and associates [4] suggests that similar increased serum levels of inflammatory mediators and increased consumption of complement and adhesion molecules occur during cardiovascular surgery in children with or without CPB. With respect to complement activation, Ascione and associates [2] concluded in their prospective randomized study between off-pump and on-pump surgery patient groups that C3a and C5a rose early after surgery in both groups when compared with baseline values. The heparin–protamine interaction is known to activate the classic complement pathway, which is also inevitable in the off-pump procedure.

Fearing hemodynamic deterioration during off-pump surgery when performing an anastomosis to the distal branch of LCX [5] and because no satisfactory conclusion regarding complement activation in the on-pump or off-pump procedure was obtained, we used the on-pump procedure by taking prophylactic measures against hemolysis.

Kypson and associates [6] reported a successful perioperative management against cardiac surgery.
associated with paroxysmal cold hemoglobinuria. Their strategy against hemolysis includes normothermic CPB without topical cooling and cold crystalloid cardioplegia which prevented the blood agglutination in coronary circulation. Pagani and associates [7] reported a case with mitral valve reconstruction in sickle cell disease and suggested the preoperative partial exchange transfusion followed by total exchange transfusion at the time of operation to reduce the level of abnormal cells during bypass. So far, we could not find any literature regarding PNH patient with cardiovascular disease which entails CPB for correction, such as valvular diseases, and we believe the present report might be a suggestion for radical repair of such patients under CPB.

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References