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Case Report
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Off-Pump Coronary Revascularization Using Bilateral Internal Thoracic Arteries in a Patient with Paroxismal Nocturnal Hemoglobinuria

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Abstract

Paroxysmal Nocturnal Haemoglobinuria (PNH), an ultra-orphan disease with a prevalence of 15.9 per million in Europe [1]. We report the first case in the literature of Off-Pump Coronary Revascularization Using Bilateral Internal Thoracic Arteries in a patient with paroxismal nocturnal hemoglobinuria.

A 36-year-old man presented to the emergency departmentfor acute non- ST elevation myocardial infarction (NSTEMI).The patient presented paroxismal nocturnal hemoglobinuria diagnosed in 2016. Coronary angiography revealed a moderate stenosesostial and proximal in the left anterior descending (LAD) coronary artery. There were chronicle stenoses of the left circumflex coronary artery (LCX) and injury of 95% of the RCA. The conduits used for coronary revascularization were both internal thoracic arteries (left ITA–right ITA [LITA-RITA]). We consider that OPCABG using BITA can be safely performed with low in-hospital mortality and complications rates, even in patient with PNH.

Keywords: Off-pump coronary revascularization; Bilateral internal thoracic arteries; Paroxismal nocturnal hemoglobinuria

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Introduction

Paroxysmal Nocturnal Haemoglobinuria (PNH), an ultra-orphan disease with a prevalence of 15.9 per million in Europe, is a life-threatening disorder, characterized by haemolysis, bone marrow failure and thrombosis [1]. PNH is based on a clonal defect of hematopoietic stem cells characterized by deficiency in glycosyl-phosphatidylinositol (GPI)-anchored surface proteins due to mutations within the X-chromosomal PIG-Agene [2]. PNH is an acquired hemolytic anemia associated with an increased risk to develop thrombocytopenia, atypical venous thrombosis and hypo plastic bone marrow [2].

We report the first case in the literature of Off-Pump Coronary Revascularization using Bilateral Internal Thoracic Arteries in a patient with paroxismal nocturnal hemoglobinuria.

Case Report

A 36-year-old man presented to the emergency department for acute non- ST elevation myocardial infarction (NSTEMI) with elevation of troponins. Previous medical history included dilated cardiomyopathy with dyspnea funtional class II-III in study and coronary disease of three vessels. In addition, the patient presented paroxismal nocturnal hemoglobinuria diagnosed in 2016 and treated with Eculizumab. On admission, laboratory tests exhibited normal kidney function (blood urea nitrogen of 29mg/dl and creatinin of 0.93 mg/dl) and normal liver function. Leucocytes 4690/mm^3. Differential blood count revealed 53.7% neutrophils, 33.7% lymphocytes, 11.1% monocytes, and 0.9% eosinophils. Hemoglobin 10.1 g/dl, Platelets 113.000/mm^3. Coronary angiography revealed a moderate stenosesostial and proximal in the left anterior descending (LAD) coronary artery. There were chronicle stenoses in the proximal segment of the left circumflex coronary artery (LCX) and injury of 95 % in the third proximal of the RCA. Echocardiography showed reduced left ventricular function (Ejection fraction: 32 %), severely dilated left ventricle and global hypokinesia. The conduits used for coronary revascularization were both internal thoracic arteries (left ITA–right ITA [LITA-RITA]). The left internal mammary artery was harvested and

anastomosed to the left anterior descending artery. The technical configuration was in-situ anastomoses of the LITA to the left anterior descending artery; and the RITA, after being divided at its origin and bifurcation, was connected end to side to the in-situ LITA as a sequential T graft to two circumflex arteries. Saphenous vein grafts were anastomosed to the posterior descending coronary artery. It was reported that skeletonized harvesting of ITA offer more conduit length and was associated with a lower incidence of sternal infection so we use this technic routinely. The procedure was off-pump without complication. The patient was transferred to the ICU after surgery. During the postoperative subcutaneous heparin 5,000 units was administered 2 times daily to avoid thromboembolism. Eculizumab (900 mg) was administrated during the postoperative to optimize hematological parameters.

Discussion

Thromboembolism is the most common cause of mortality in PNH, and the 4-year survival of patients presenting with thrombosis at diagnosis was only 40% before the era of eculizumab [3]. Eculizumab is a humanized monoclonal antibody that blocks terminal complement pathway by binding to C5. This drug has dramatically changed the natural history of PNH. Eculizumab increases transfusion independency, reduces the risk of further thrombotic events and also health-related quality of life. [4].

Cardiac surgery in PNH patients is associated with several possible complications. PNH-induced granulocytopenia increases the risk of postoperative infection. The aggravation of hemolysis by extracorporeal circulation in cardiac surgery due to complement activation from either contact of blood with the foreign material surfaces during cardiopulmonary bypass circuit, or use of protamine to neutralize systemic heparin after cardiopulmonary bypass and tissue injury is well known [2]. We consider that OPCABG using BITA can be safely performed with low in-hospital mortality and complications rates, even in patient with PNH. Surgical tricks and the new technology in coronary stabilizers allow surgeons to perform a complete myocardial coronary revascularization using the best available arterial conduit (BITA) [5]. There have been 5 cases reported previously of patients with PNH undergoing cardiac surgery, but this is the first case in where we combine Off-Pump Coronary Revascularization Using Bilateral Internal Thoracic Arteries in a patient with paroxismal nocturnal hemoglobinuria.

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