Urgent Off-Pump Coronary Bypass Grafting in Moyamoya Disease: A Case Report

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ABSTRACT

Moyamoya disease is a rare progressive cerebrovascular occlusive disease that predominantly occurs in countries in Northeast Asia. We report a 54-year-old male patient with moyamoya and coronary artery disease, on whom urgent off-pump coronary artery bypass grafting was performed after cardiopulmonary reanimation due to ventricular fibrillation. This patient had left main trunk disease, a Pudenz peritoneal catheter (Medtronic, Inc., Minneapolis, MN, USA) for liquor drainage, bilateral internal carotid artery occlusion, diffuse peripheral atherosclerotic artery disease, and chronic renal insufficiency. We prefer off-pump surgery to avoid the risk of intraoperative hypotension and minimize the risk of perioperative cerebral ischemic complications.

INTRODUCTION

Moyamoya disease (MMD) is a chronic occlusive cerebrovascular disease characterized by steno-occlusive changes of the internal carotid artery (ICA) and the vascular network at the base of the brain. At the same time, tiny blood vessels (“moyamoya” vessels) at the base of the brain open up in an apparent attempt to supply blood to the area distal of the blockage. In Japanese, “moyamoya” is an “expression for something hazy just like a puff of cigarette smoke drifting in the air” [Suzuki 1969]. MMD was first described by J. Takeuchi and K. Shimizu in 1969 [Suzuki 1969]. Ten to 15 percent of the cases of this disease are inherited. The risk of having MMD in family members is about 30 to 40 times higher than in the general population. The disease is most common in Northeast Asia, where the incidence is approximately 0.35 per 100,000. In Europe and the United States, the percentage is much smaller, that is, 0.086 people per 100,000. The disease usually occurs in children under 10 years of age or in people at 30 to 40 years of age. Women are more likely to have this disease with a 1.8:1 ratio [Kim 2016].

We report a case of a moyamoya patient with unstable angina pectoris with left main trunk disease, frequent transient ischemic attacks (TIAs), and ventricular fibrillation (VF), with cardiopulmonary reanimation before urgent off-pump coronary artery bypass grafting (CABG) surgery. Our main concern was brain protection during off-pump CABG surgery because of insufficient cerebral perfusion due to bilateral ICA occlusion.

CASE REPORT

A 54-year-old male patient was admitted to our hospital due to sudden chest pain. In his medical history, he had MMD disease, which was diagnosed when he was 25 years old after he had suffered ischemic brain stroke. A Pudenz peritoneal catheter (Medtronic, Inc., Minneapolis, MN, USA) was implanted 1 year after the stroke. MMD was diagnosed on the basis of the subsequent cranial computed tomography (CT) angiography, which showed a typically abnormal vascular network with the occlusion of both ICAs. He also had insulin-dependent diabetes mellitus, arterial hypertension,
diffuse peripheral atherosclerotic artery disease diagnosed with CT angiography, hyperlipidemia, and chronic renal insufficiency. Coronary angiography was performed on the same day and showed triple vessel disease with left main trunk disease, proximal diagonal stenosis, proximal stenosis of right coronary artery, and chronic occlusion of the circumflex artery with no collateral flow (Figure).

Transesophageal ultrasound showed a dilated left ventricle chamber with lower ejection fraction of 39% with a kinetic lateral wall of the left ventricle chamber. On the third day, he had VF with cardiopulmonary reanimation and multiple ventricle defibrillations. After reanimation, he was intubated and mechanically ventilated with normal arterial pressure, and with no changes in electrocardiogram (ECG). After that, we performed urgent off-pump CABG, and the heart was stabilized by using Octopus (Medtronic) tissue stabilizers. The left internal mammary graft was anastomosed to the left anterior descending artery, which was calcified in the entire proximal part, and the saphenous vein graft was anastomosed to the right coronary artery and the diagonal branch separately without any intraoperative complications. During the operation, we constantly paid attention to systolic blood pressure and blood PaCO$_2$, which was above 100 mmHg and 40 mmHg. The patient was transferred to the cardiovascular intensive care unit, where on the second and the third days he had self-limiting ventricular tachycardia. On the ninth day, an implantable cardioverter defibrillator (ICD) (Medtronic Maximo; Medtronic, Tolochenaz, Switzerland) was implanted through the left cephalic vein. After that, he was discharged from the hospital on the 11th day after surgery.

**DISCUSSION**

To the best of our knowledge, there are 6 MMD patients in Croatia, and this is the second case of a white patient with MMD operated on in Europe and the fifth in the world, especially if we take into consideration that it is the first case of urgent off-pump CABG performed after cardiopulmonary reanimation in an MMD patient. In the first case, a 56-year-old Japanese female patient had the left anterior descending artery treated by balloon angioplasty and followed by elective left anterior mini-thoracotomy and minimally invasive direct CABG. The second case was a 56-year-old Korean female patient in whom triple off-pump CABG was performed [Kim 2007]. The third case was a 40-year-old Japanese woman with left main trunk disease in whom off-pump CABG was performed, supported by intra-aortic balloon pumping to maintain appropriate blood flow intraoperatively [Okamoto 2010]. In the last case, a 53-year-old Turkish male was treated with off-pump CABG [Coşkun 2018].

We cannot be sure whether coronary artery disease in our patient was caused by MMD or the cause was (or were) insulin-dependent diabetes mellitus, arterial hypertension, diffuse peripheral atherosclerotic artery disease, and/or hyperlipidemia. After surgery, brain CT scans were performed and did not show any ischemia or brain hemorrhage.

In our case, we opted for urgent off-pump CABG because after reanimation the patient was hemodynamically stable without ECG abnormalities. The decision to perform off-pump CABG surgery was made to reduce the risk of intraoperative hypotension to minimize the risk of perioperative cerebral ischemic complications, as we believe that off-pump CABG provides better brain protection. We were prepared to use the heart-line machine if necessary. In conclusion, we believe that off-pump CABG can be performed after reanimation of unstable patients with MMD, with special care to be taken for arterial pressure and PaCO$_2$.

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