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Coronary bypass surgery in essential thrombocytemia

Esansiyel trombositemide koroner baypas cerrahisi

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Introduction

Essential thrombocytemia (ET) is a rare myeloproliferative disease characterized by a persistent increase in platelet numbers and platelet dysfunction. The disease manifests clinically by hemorrhage and/or thromboembolic complications. Coronary artery involvement is more prevalent than previously thought but there are only 9 cases of coronary bypass published in the literature. We present a case of essential thrombocytemia presenting with an acute coronary syndrome treated surgically employing the off-pump beating heart surgery.

Case report

A 61-year-old man with an established diagnosis of ET was referred to our hospital for rapidly progressing episodes of chest pain. On admission, he was in a stable hemodynamic condition. Laboratory investigation revealed a hemoglobin of 8.9 g/dl, Hct - 30% and a platelet count of 930 x 10^9 /L. Electrocardiogram (ECG), creatine kinase-myocardial band isoenzyme and troponin levels were normal. Inherited thrombophilia tests (Protein C, Protein S, antithrombin III deficiency, Factor V Leiden, prothrombin 20210 A, MTHFR mutations, antiphospholipid antibodies) were normal. Following an initial medical treatment with nitrates, beta-blockers, angiotensin converting enzyme inhibitors, aspirin, statins, sedatives and continuing anti-platelet therapy with Anagrelide (Thromboreductin™ - AOP Orphan Pharmaceuticals, Austria) a coronary angiogram was performed, revealing an irregular 90% stenotic lesion of the proximal left anterior descending (LAD) and first diagonal coronary arteries. The other coronary arteries were disease-free. A coincidental finding during the angiogram was segmental total occlusion of the right external iliac artery with extensive collateral vessel development filling the femoral arteries. The coronary lesion was considered unsuitable for an angioplasty and a decision for surgical therapy was made. The platelet count was found to be high (851 x 109 /L) despite therapy so the patient was maintained on IV heparin and nitroglycerin infusions to relieve the unstable angina while an increased dose of anti-platelet therapy with Anagrelide (2 mg/day) was administered until the platelet count was reduced to 326 x 10^9 /L.

At this point; hemostasis parameters and plasma Von Willebrand factor (VWF) levels were normal but epinephrine-induced secondary platelet aggregation was inhibited.

In view of the deleterious effects of cardiopulmonary bypass (CPB) and the anemia and history of gastrointestinal bleeding of the patient we chose to perform the operation with the off-pump, beating heart method. A single internal mammary artery graft (IMA) was used to bypass the lesion of the LAD coronary artery. The quality of the IMA graft, LAD and anastomosis was excellent and the operation was completed without complications. The patient was transferred to the intensive care unit in a stable condition requiring no inotropic support.

During the operation the patient was anticoagulated with heparin (5mg/kg) and the activated clotting time (ACT) was maintained at 350 sec during occlusion of the LAD and IMA. After completion of the anastomosis the heparinization was partially reversed with protamine sulphate and the operation was completed with the ACT at 150 sec.

The early postoperative course was uneventful. All hemodynamic parameters were stable requiring no inotropic support. The ECG was normal and the first postoperative hemogram revealed a Hb of 9.1 g/dl, Hct of 32% and a platelet count of 262 x 10^9/L. Chest tube drainage was 300 cc in the first two hours with a slight increase of 200 cc in the third hour. The ACT at this time was 126 seconds, the platelet count was 232 x 10^9/L and hemostasis parameters were normal. Chest tube drainage loss was replaced with packed red cells.

On the fifth postoperative hour, the patient was hemodynamically stable, awake and being weaned off the ventilator when he suddenly went into ventricular fibrillation which was reverted to normal rhythm and hemodynamic state with a brief period of cardiac resuscitation. A few minutes later a second cardiac arrest accompanied by a long period of generalized convulsions occurred and the patient expired despite all efforts.

Discussion

Essential thrombocytemia is a rare, clonal chronic myeloproliferative disease of unknown origin characterized by a persistent increase in platelet numbers (>600x10^9/L) and platelet dysfunction. Originally described as a hemorrhagic disorder, recent studies have shown it is manifested with thrombotic and thromboembolic complications. The incidence of thrombotic and hemorrhagic complications and the overall rates of thrombosis and hemorrhage at diagnosis range from 9% to 84% and from 3.9% to 63%, respectively. Age, a previous thrombotic event and long duration of thrombocytosis were identified as major risk factors for thrombosis in the controlled series. Recent studies have suggested that vascular complications can also be predicted from the biological characteristics of the disease, markers of hypercoagulability (thrombophilia, Factor V Leiden, antiphospholipid antibodies) and general cardiovascular risk factors (hypertension, hypercholesterolemia, smoking). Paradoxically, a high platelet count (≥1500x10^9/L) is a major predictor of bleeding rather than thrombotic complications. Normalization of the platelet count is accompanied by restoration of a normal plasma VWF multimeric distribution .

Patients with a high risk of thromboembolic complications or bleeding are candidates for interventions to lower platelet counts. Several agents are now available for initial treatment of ET to prevent these complications. Anagrelide and Hydroxyurea are effective in reducing high platelet counts in ET.

Platelets have a well-defined critical role in coronary thrombosis. The incidence of coronary artery disease in ET has been reported as 9.4 % in patients 40 years and older with a high incidence of acute myocardial infarction (AMI) (1). In addition, many patients with ET initially present with symptoms related to small or large vessel thrombosis.

Experience in coronary revascularization in patients with ET is very limited. Our search of the medical literature revealed only nine cases of ET treated surgically for acute coronary syndromes (2-10).

All cases were operated utilizing cardiopulmonary bypass. Six of the studies report early successful outcomes (2-4, 6-8). The long-term results of surgery however are not available. One patient suffered an AMI and cerebral infarct in the early postoperative period (5)

One patient died in the postoperative sixth month, again due to an AMI and cerebrovascular accident (CVA) (8). Another case, a 26-year-old previously healthy man suffered an AMI and despite coronary artery

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bypass grafting (CABG) required left ventricular assist device (LVAD) implantation due to deterioration of hemodynamic state. The patient was later transported to specialized center with LVAD support and eventually underwent successful heart transplantation (10). Three patients presented clinically with AMI (2, 9, 10) and two others had history or evidence of peripheral artery thrombosis; one of the common iliac artery and the other of the carotid artery (4, 7). Surprisingly none of the cases have encountered any problems regarding hemorrhage postoperatively. One would expect that the deleterious effects of CPB would aggravate the inherent tendencies of bleeding in ET. Only one of these patients required blood transfusions for hematoma of the groin after a coronary angiogram (7).

The limited surgical experience with ET demonstrates that this entity carries a high rate of morbidity and mortality mainly due to thromboembolic complications despite the meticulous efforts to monitor and manipulate platelet numbers and function.

Reports of successful surgery stress the importance of a combination therapy with cytoreductive agents and antiaggregants to prevent thromboembolic events (6) There is, however a delicate balance of hemorrhage versus thromboembolism to be considered in the case of surgical interventions. Platelet functions are very unpredictable in ET causing hemorrhage and thrombosis in the same patient during the same time frame. Cardiopulmonary bypass causes severe reductions in platelet numbers and function (aggregation) alongside depletion of coagulation factors rendering the patient prone to postoperative bleeding. Theoretically the combination of CPB and an ET patient with sub-normal platelet functions is a potential hematological disaster. Keeping this in mind, we decided to utilize the off-pump beating heart method for revascularization. It is clear in this case that in an attempt to avoid the possible hemorrhagic complications of ET and CPB we have run straight into another devastating (thrombotic) complication. Despite our efforts to normalize platelet numbers preoperatively with Anagrelide and maintain an acceptable level of anticoagulation perioperatively the patient suffered a massive thromboembolic episode. Although an autopsy was not available we postulate that this thromboembolic episode must have happened almost instantaneously because at no stage were there any indications of a problem, such as deteriorating hemodynamics, change in ECG or consciousness.

The lack of response to all resuscitation efforts and the development of cardiac standstill in a matter of minutes alongside convulsions during cardiac arrest suggests a massive thrombosis of not only the bypass vessel but probably all the coronary arteries and major cerebral vessels.

A number of studies have demonstrated that platelet function tests can predict 'major adverse cardiac events' in cardiovascular disease but none of these assays have yet been sufficiently studied in large clinical trials to become part of standard clinical evaluation . Further research is necessary to understand the unpredictable nature of platelet function in patients with ET undergoing major surgical procedures.

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Two-stage successful surgery in an aortic coarctation case operated initially for ascending aortic aneurysm

Öncelikli olarak assandan aort anevrizmasına müdahale edilen aort koarktasyonlu bir olguda iki evreli başarılı cerrahi tedavi

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Introduction

Aortic coarctation is a serious pathology required surgical treatment. About 50% of uncorrected isolated aortic coarctation cases are lost up to the age of 10, only 10% may reach the age of 50 (1). The most common reason for death from untreated aortic coarctation is the aneurysm or rupture of aorta or side branches with a rate of 23% (1). Aortic insufficiency resulting from annuloaortic ectasia and ascending aortic aneurysm together with aortic coarctation rarely occur, and surgical treatment is difficult. It is very important to decide whether surgical operation will be of one and two stage, and to determine intraoperative strategy.

Aortic coarctation is a congenital vessel disease that can cause such complications as myocardial infarction, congestive cardiac failure,

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