CASE REPORT

Syndrome of Thrombotic Microangiopathy in A Patient Submitted to Mitral Valve Exchange with Biological Prosthesis Implant and Extracorporeal Circulation: Case Report

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Abstract

Introduction: The development of cardiopulmonary bypass (CPB) has provided advances in cardiovascular surgery techniques. Considering its mechanism of action, as a non-endothelial surface, mechanical hemolysis is expected, but a complete investigation of the condition is essential to rule out other differential diagnoses.

Case Report: A 34-year-old female patient underwent mitral valve replacement surgery with a biological prosthesis implant and evolved in the postoperative period with microangiopathic hemolysis associated with renal injury, favoring the diagnostic hypothesis of Hemolytic Uremic Syndrome (aHUS).

Discussion: Although mechanical hemolysis is expected in the evolution of patients undergoing CPB, the correct investigation and microscopic review of cases is essential, avoiding underdiagnosis of serious etiologies, such as thrombotic microangiopathies. This report highlights the importance of a careful investigation and microscopic correlation in hemolytic anemias due to their diverse etiologies.

Keywords: Cardiovascular Surgery, Extracorporeal Circulation, Microangiopathic Hemolytic Anemia, Intravascular Hemolysis, Mechanical Hemolysis.

1 | INTRODUCTION

Extracorporeal Circulation (ECC) was applied for the first time in 1953, in a correction of interatrial septum defect and since then generated advances in cardiovascular surgery techniques (1). Considering its mechanism of action, involving a non-endothelial surface, it is known that ECC often generates some negative effects, such as severe systemic inflammatory response with mobilization of the leukocyte margin pool, hemodilution and mechanical erythrocyte injury, causing mechanical intravascular hemolytic anemia (2).
Thus, patients undergoing this type of procedure are expected to evolve with hemolysis markers of mechanical origin. However, considering the different etiologies of hemolysis, it is essential to rule out other differential diagnoses. For this, a basic evaluation including microscopic analysis of the peripheral blood smear (which is expected to show polychromasia, anisocytosis and schizocytes in cases of mechanical hemolysis), hemoglobin, reticulocyte count, bilirubin, direct coombs and haptoglobin levels can be very helpful (3).

Among hemolytic anemias etiologies, thrombotic microangiopathies (TMA) are of great importance, corresponding to a set of clinical entities that share morphological changes and the triad of microangiopathic hemolytic anemia, thrombocytopenia and organic dysfunction secondary to vascular occlusion caused by microvascular thrombi (4). Causes of TMA include infections (by diverse agents as HIV, Shigella dysenteriae, E. coli or Streptococcus pneumoniae), systemic diseases (malignant hypertension, vasculitis, systemic lupus erythematosus, scleroderma, antiphospholipid antibody syndrome, disseminated intravascular coagulation and neoplasms), drugs (such as calcineurin inhibitors, P2Y12 antagonists), transplantation, pregnancy, metabolic or cell signaling changes (DKGE mutations and cobalamin C) or genetic/acquired changes that leads to ADAMTS13 (ADAM Metallo peptidase With Thrombospondin Type 1 Motif 13) deficiency and abnormalities in complement system (5).

Due to the multiplicity of differential diagnoses in the context of hemolytic anemias, we highlight, through this report, the importance of correct investigation and evaluation of hemolysis after cardiovascular surgery with ECC, emphasizing thrombotic microangiopathies as a severe condition that requests rapid diagnosis and intervention. (6)

2 | CASE REPORT

A 34-year-old female was admitted to the Intensive Care Unit in the postoperative period of mitral valve replacement for rheumatic valve disease, with implantation of biological prosthesis number 27 and excision of a thrombus in the left atrium, submitted to extracorporeal circulation for 125 minutes.

Previously healthy, she evolved in the postoperative period with thrombotic microangiopathy syndrome (TMA). TMA was quickly suggested and supported by the following laboratory changes: DHL 2669 U/L (VR 207 to 414 U/L), mean of two schizocytes per high power field in peripheral blood smear, hyperbilirubinemia with a predominance of indirect bilirubin, decreased platelet count compared to baseline, metabolic acidosis and worsening of renal function. Rapid clinical and metabolic support was offered and atypical Hemolytic Uremic Syndrome (aHUS) was considered the main diagnostic hypothesis, given the evident renal impairment. Eculizumab (inhibitor of C5 complement factor) was considered, but she evolved with rapid clinical deterioration and death, despite the established procedures.

Clinical context strongly suggested the hypothesis of aHUS, especially because it is a TMA with impaired renal function. Complementary exams were performed to exclude other diagnoses, such as Anti-Nuclear Factor HEP2 to exclude Systemic Lupus Erythematosus. Seric complement was consumed, with C3 dosage of 35 mg/dl (reference range: 90 to 170 mg/dl), C4 of 10 mg/dl (reference range 12 to 36 mg/dl) and CH 50 of 34 u/CAE (reference value ≥60 u/CAE), reinforcing the hypothesis of aHUS. It was not possible to rule out thrombotic thrombocytopenic purpura, as the patient died before ADAMTS 13 dosage, and also was not possible to perform a renal biopsy due to the rapid evolution and clinical deterioration.

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Patients undergoing cardiovascular surgery with extracorporeal circulation commonly evolve to some degree of mechanical (intravascular) hemolysis. The contact of red blood cells with a non-endothelial surface associated with cell membrane fragility, promotes direct injury. Some factors, such as prolonged ECC period, characteristics of circuits (roller pump or centrifugal pump) and their calibration can also influence this injury. In addition, excessive intraoperative bleeding with massive aspiration can contribute to increase intravascular hemolysis, both due to the role of non-endothelial circuits that culminate in erythrocyte injury and due to blood transfusions, which contains more hemolyzed red cells as a result of the duration of storage and handling of the unit itself. (7)

However, it is important to emphasize that there are other etiologies of hemolysis in this context, and consider differential diagnosis as drug hemolysis, immune-mediated hemolysis (by irregular antibodies or auto-antibodies), among others. Given the severity of TMA, its immediate recognition is essential, especially in patients with unusual or more severe clinical evolution. (8)

The evolution to TMA after cardiovascular surgery is multi-factorial, and some associated factors appear to be the inactivation of ADAMTS 13 as a consequence of the inflammatory process and the procedure itself, promoting greater amounts of high molecular weight multimers of Von Willebrand factor and ADAMTS 13 reduced action (9). In the context of heart transplantation, for example, a common cause is the use of calcineurin inhibiting medications (10). In myocardial revascularization surgeries, endothelial injury was considered as a trigger for TMA development (9). In addition, genetic predisposition seems to influence the evolution and mutations in several coding regions of complement system have already been reported (11). In valve replacement procedures, there appears to be stimulation of the complement system both by the procedure itself and by the use of extracorporeal circuits (12). We also reinforce through this report the importance of microscopic review of peripheral blood smear slides of patients undergoing cardiovascular surgery with ECC, emphasizing the importance of microscopic findings as an alert for possible underlying diseases or atypical manifestations. (13, 14)

REFERENCES


