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ANESTHESIA FOR MITRAL VALVE REPLACEMENT IN A PATIENT WITH BETA THALASSEMIA MAJOR : CASE REPORT

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ABSTRACT

Post-spinal Thalassemias constitute a heterogeneous group of inherited genetic disorders characterized by deficient synthesis of hemoglobin polypeptide chains, with beta-thalassemia being the most prevalent autosomal recessive hereditary hematologic disease. Caused by mutations in the beta-globin gene on chromosome 11, it leads to anemia with symptoms such as fatigue, dyspnea, weakness, headaches, and hepatosplenomegaly. Peripheral blood smears typically reveal anisocytosis, poikilocytosis, and numerous nucleated erythrocytes. The standard treatment involves lifelong regular blood transfusions and chelation therapy to manage iron overload. This report details the anesthetic and surgical management of a complex patient with beta-thalassemia major undergoing mitral valve replacement. The patient, who also presented with hypertension, diabetes, hypothyroidism, pulmonary hypertension, gastroesophageal reflux disease, and generalized anxiety disorder, received multimodal balanced general anesthesia combined with interfascial blockade of the serratus and deep pectoralis muscles. A minimally invasive biological mitral valve replacement was successfully performed. Postoperatively, the patient exhibited stable hemodynamic and ventilatory parameters and was extubated in the operating room without complications. The discussion emphasizes the importance of regular serum ferritin monitoring in transfusion-dependent patients and the necessity of meticulous perioperative care to prevent complications such as hypoxemia and hypovolemia, particularly given the risk of erythrocyte sickling in this population.

Keywords: Beta-Thalassemia, Thoracic Surgery, Mitral valve, Anemia, Pulmonary hypertension.

INTRODUCTION

Thalassemias comprise a heterogeneous class of inherited genetic conditions defined by a quantitative deficiency in the production of one or more polypeptide chains constituting the hemoglobin molecule.[1,2] Beta-thalassemia is recognized as the most prevalent autosomal recessive hereditary hematologic disorder among various populations and results from mutations in the β-globin gene, located on chromosome. [11] To date, more than 350 distinct mutations responsible

for this condition have been identified, with most representing point mutations situated within functionally significant regions associated with the gene expression of the globin protein.[3]

In the context of β-thalassemia, a reduced synthesis of the beta-globin chain leads to a relative excess of the alpha-globin chain, another essential component of hemoglobin. Consequently, these excess alpha-globin chains aggregate, resulting in damage to red blood cells and subsequent hemolysis. Clinically, individuals with β-thalassemia present with anemia and its associated sequelae, including fatigue, dyspnea, weakness, and headaches, potentially extending to hepatosplenomegaly, growth retardation, cardiac complications, pancreatic alterations, and diabetes.[4,5] The clinical manifestations observed in β-thalassemia major (BTM) are directly influenced by two main sets of factors:

1) the severity of chronic hemolysis, the body's capacity to compensate for anemia, the effects of chronic hypoxia, splenomegaly, and skeletal changes. Consequently, more effective transfusion regimens are associated with a lower risk and reduced severity of these complications;

2) the risks associated with red blood cell transfusions, such as transmission of infectious agents and alloimmunization, but critically, complications arising from iron overload. The latter depends on the efficacy and adherence to iron chelation therapy, aiming to maintain a negative iron balance, mitigating the adverse effects associated with excessive use of chelating agents.[6]

Consequently, the initial suspicion of β-thalassemia minor often arises from a complete blood count indicating microcytic and hypochromic anemia. The characteristic peripheral blood smear in the major form shows anisocytosis, poikilocytosis, and a substantial number of nucleated erythrocytes. Definitive diagnosis requires hemoglobin electrophoresis and genetic analysis (DNA sequencing) of the parents, confirming their mandatory carrier status for a thalassemia gene.[7] Individuals who present with severe anemia in early childhood, requiring regular blood transfusions from that point onward, are classified as carriers of β-thalassemia major (BTM).[8] Consequently, treatment usually involves a lifelong regimen of regular blood transfusions and chelation therapy to control iron overload. Hematopoietic stem cell transplantation from healthy donors represents a potentially curative option, although its application is limited due to the substantial risks associated with the procedure and its high cost.[9] Inadequate treatment and the inherent pathophysiology of BTM are strongly correlated with a higher mortality rate in affected individuals, with specific comorbidities occurring more frequently, including heart disease, infections, liver disease, malignancy, thromboembolism, and severe anemia.[10]

Considering the rarity of the disease along with the specific surgical procedure performed on the patient in this report, the present study aims to document the anesthetic and surgical management employed, as well as to synthesize relevant data from the existing literature regarding the anesthetic management of patients with β-thalassemia major (BTM) in various surgical scenarios.

CASE REPORT

A 43-year-old female patient, measuring 164 cm in height and weighing 79 kg, with a medical history of hypertension, diabetes mellitus, hypothyroidism, pulmonary hypertension, gastroesophageal reflux disease, beta-thalassemia major, and generalized anxiety disorder, underwent mitral valve replacement on October 8, 2024, at Hospital Encore in Aparecida de Goiânia, Brazil. The patient reported a family history of thalassemia trait in both parents and has been receiving blood

transfusions every two weeks since birth. At age 20, she experienced an iron overload crisis that required hospitalization due to pleural effusion.

Regarding the surgery, initial monitoring included electrocardiography, pulse oximetry, and non-invasive blood pressure measurement. Venous access was established with a 16G intravenous catheter in the right upper limb and a 14G intravenous catheter in the left upper limb. After light sedation, the left radial artery was cannulated under local anesthesia using the Seldinger technique. Depth of anesthesia was monitored using a processed electroencephalogram index (CONOX™), and transesophageal echocardiography was employed throughout the procedure.

The anesthetic technique employed was balanced multimodal general anesthesia combined with interfascial plane blocks of the serratus anterior and pectoralis major muscles. The patient underwent biological mitral valve replacement via a minimally invasive technique involving a right anterolateral minithoracotomy. This approach involved two points of cardiac access: one through the right midclavicular line at the fourth or fifth intercostal space (periareolar) and the other through the anterior axillary line. After skin incision and dissection of the subcutaneous tissue, Surgisleeve™ retractors were placed, and pericardiotomy was performed under direct visualization to access the cardiac structures. Subsequently, cardiopulmonary bypass (CPB) was established with full heparinization, cannulating the right femoral artery and vein. A left atriotomy was then performed, followed by dissection and removal of the thickened mitral valve with restricted opening. A long-lasting biological mitral valve prosthesis (No. 31 – Medtronic – Hancock II) was implanted and secured to adjacent structures, followed by closure of the left atrium. The patient was weaned from CPB after a total duration of 102 minutes, with an aortic cross-clamp time of 78 minutes. Right thoracic drainage was performed using a Blake drain – 24Fr, and the procedure was completed with layered closure of the subcutaneous tissue and skin. Postoperative echocardiography demonstrated good function of the implanted prosthesis and normal left ventricular function.

After the completion of the surgery, the patient maintained stable hemodynamic and ventilatory parameters, allowing extubation in the operating room without adverse events. The patient was transferred to the intensive care unit (ICU) breathing spontaneously with supplemental oxygen via nasal cannula at 3 L/min and without the need for vasoactive drugs. The patient remained in the ICU for two days without complications related to the surgical procedure or anesthesia, and was subsequently transferred to a general ward. No blood transfusions were required during the hospital stay. The patient was discharged for outpatient follow-up after a total hospital stay of 3 days, without postoperative complications and with good clinical progress, continuing her usual medications for underlying conditions and with instructions on postoperative care.

DISCUSSION

Placental The existing body of literature contains a limited number of reports detailing anesthetic management strategies across various surgical procedures. Furthermore, the occurrence of complications and the need for perioperative blood transfusions are infrequently documented in research, thus representing a considerable challenge for anesthesiologists. Olivieri et al., in their study involving patients with β-thalassemia major (BTM), demonstrated that for an individual patient, a prolonged period during which ferritin levels remain below 2,500 ng/ml correlates with a lower likelihood of developing cardiac disease and a higher overall survival rate (90% at 15 years)[11]. Consequently, periodic assessment (every two to three months) of serum ferritin levels

is considered essential for the longitudinal monitoring of patients undergoing regular red blood cell transfusions.[11] The investigation by Angulo et al. emphasizes the importance of quantifying hepatic and myocardial iron deposition through magnetic resonance imaging, and correlating this methodological approach with serum ferritin levels to stratify the degree of iron overload, thus guiding subsequent therapeutic interventions based on these findings.[12]

In the present clinical case, the patient underwent a comprehensive panel of laboratory investigations, including measurement of ferritin levels, transthoracic and transesophageal echocardiography, and received specialized consultations from a nephrologist and a hematologist. Given the patient's multiple pre-existing comorbidities, a standardized anesthetic protocol was selected and implemented by the surgical team. The intraoperative phase proceeded uneventfully, with the patient requiring no blood transfusions and showing no clinical or laboratory evidence of hemolysis. Throughout the surgical procedure, meticulous attention was directed toward the prevention of hypoxia, along with vigilant monitoring for potential cardiac complications in the postoperative period. Recent findings reported by Borgna-Pignatti et al. indicated improved survival outcomes in Italian patients with β-thalassemia major, potentially attributable to more effective assessments of iron overload, including myocardial evaluation using T2* magnetic resonance imaging, and the use of oral iron chelators, particularly combination therapy involving deferoxamine and deferiprone. This therapeutic regimen appears to offer greater cardioprotection, resulting in a lower incidence of cardiac complications and reduced mortality.[13]

According to Baş and Özlü, the administration of general anesthesia and the physiological stress associated with surgical trauma may increase the risk of complications due to alterations in core body temperature, blood pH, oxygen tension, and intravascular fluid volume. Consequently, diligent perioperative care and postoperative monitoring are crucial to prevent hypoxemia, hypovolemia, hypothermia, acidosis, and the need for increased blood transfusions, particularly considering the propensity for erythrocyte sickling in this patient population.[14]

CONCLUSION

Patients with β-thalassemia major requiring surgical intervention demand meticulous and individualized care, beginning with a comprehensive pre-anesthetic evaluation and extending through to patient discharge. Tailoring the management strategy to address comorbidities beyond thalassemia is essential for the safe and successful conduct of any surgical procedure.

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EDITORIAL AND REVIEW

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