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Initial Thrombocyte Concentrate Transfusion in Woman with Chronic Immune Thrombocytopenia Purpura (ITP) Who Underwent Mitral Valve Replacement Surgery: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 42-year-old
Final Diagnosis: Post mitral valve replacement
Symptoms: Shortness of breath
Clinical Procedure: Mitral valve replacement
Specialty: Cardiac surgery

Objective: Rare coexistence of disease or pathology

Background: Chronic immune thrombocytopenia purpura (ITP) is associated with a higher incidence of adverse outcomes, increased morbidity and mortality rates, and higher health care costs, especially in open-heart surgery. The information regarding managing chronic ITP in patients undergoing mitral valve replacement (MVR) surgery is scarce, and reported cases are limited.

Case Report: A 42-year-old woman with more than 20 years of history of immune thrombocytopenia purpura (ITP) had episodes of breathing difficulties in the last 4 years. The patient was diagnosed with severe mitral stenosis (MS) and moderate mitral regurgitation (MR). Laboratory examination before surgery showed thrombocytopenia (49 000/µL). Therefore, the surgery was postponed until the platelet count exceeded 100 000/µL. The patient was given 10 units of thrombocyte concentrate 1 day before surgery and 500 mg of methylprednisolone 3 times a day orally for 5 days as preoperative management. Under a total cardiopulmonary bypass, MVR was performed using a bioprosthetic valve. Postoperative transthoracic echocardiography (TTE) showed no valvular leakage in the surrounding of the prosthetic valve and that the valve was functioning normally. Platelet monitoring was conducted, and the platelet count increased to 147 000/µL on the third day.

Conclusions: Our case report shows that aggressive preoperative platelet count correction and treatment may lower the risk associated with a low and unstable platelet count and reduce the risk of mortality and morbidity in patients with ITP who undergo MVR procedures.

Keywords: Heart Valve Prosthesis Implantation • Mitral Valve Insufficiency • Mitral Valve Stenosis • Thrombocytopenia

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Background

Chronic immune thrombocytopenia purpura (ITP) is associated with a higher incidence of adverse outcomes, increased morbidity and mortality rates, and higher health care costs, especially in open-heart surgery. The information regarding managing chronic ITP in patients undergoing mitral valve replacement (MVR) surgery is scarce, and reported cases are limited.

Case Report

A 42-year-old woman with more than 20 years of history of immune thrombocytopenia purpura (ITP) routinely visited an internal medicine specialist and received thrombocyte concentrate (TC) transfusion frequently. In the last 4 years, she had episodes of breathing difficulties after daily activities. She was diagnosed with ITP when she came into the internal medicine clinic with red-bluish spots all over her body and a low platelet count; ever since then, she has routinely visited the internal medicine specialists to manage her ITP and received thrombocyte concentrate (TC) transfusion on an as-needed basis. She denied a history of chest pain, leg swelling, or spontaneous bleeding. There was no history of hypertension, diabetes mellitus, stroke, or other comorbidities. Transthoracic echocardiography (TTE) examination 3 months before admission showed the appearance of severe mitral stenosis (MS), moderate mitral regurgitation (MR) due to rheumatic heart disease (RHD), mild tricuspid regurgitation (TR) with a high probability of pulmonary hypertension, global normokinetic, good right ventricle (RV) contractility, and no thrombus. She was diagnosed with severe MS, moderate MR, and ITP. It was decided that she would undergo mitral valve replacement (MVR) therapy.

Laboratory examination before surgery showed thrombocytopenia (49 000/µL); therefore, the surgery was postponed until the platelet count exceeded 100 000/µL. She was given 10 units of thrombocyte concentrate 1 day before surgery and 500 mg of methylprednisolone 3 times a day orally for 5 days as preoperative management. She also continued her previous daily pharmacological treatment, including bisoprolol, furosemide, spironolactone, and digoxin. After receiving the previously mentioned treatment, she underwent preoperative re-examination. There were no remarkable differences in the patient's signs and symptoms between the first and second examinations, except that the platelet count had increased to 107 000/µL. Therefore, she was now eligible to undergo mitral valve replacement surgery. However, preoperative transesophageal echocardiography (TEE) found a thrombus in the left atrium (**Figure 1**). The other TEE findings were similar to the previously performed TTE.

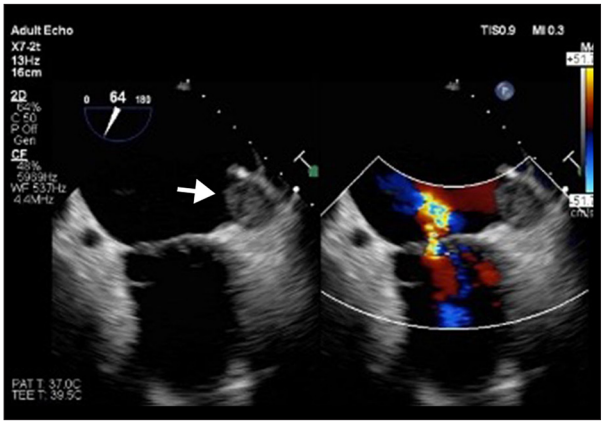

Figure 1. Thrombus found in the left atrium during preoperative TEE.

Median sternotomy and pericardiotomy were performed, followed by a total bypass with a cardiopulmonary bypass machine. The surgeon opened the right atrium (RA), followed by opening and widening the intra-atrial septal (IAS). The findings agreed with the previous TEE. A thrombus was found in the left atrium (LA) and was evacuated. (**Figure 2A**). Left atrial appendage (LAA) exclusion was performed. The mitral valve (MV) was examined, and was found to be consistent with the preoperative TTE results (**Figures 2B, 3A**). MVR was performed using a 27-mm bioprosthetic valve, which was secured with 14 sutures of siliconized braided polyester pledget Teflon. After the MV was re-evaluated to ensure the prosthetic valve was placed correctly, the IAS was closed. The patient's temperature returned to normal, left heart de-airing was performed, and the cross-clamp was removed. Atrial fibrillation with normal ventricular response was found. The RA was closed, and the patient was weaned from cardiopulmonary bypass.

Postoperative TTE showed no valvular leakage in the surroundings of the prosthetic valve, and the valve's function was normal. The TTE also showed mild TR and good ventricle contractility (**Figure 3B**). Protamine was given, IVC and SVC cannulas were removed, and the aortic cannula was removed. Two pacemakers were placed on the RV. Bleeding was 1000 cc. The patient was given 195 cc of TC and 320 cc of FFP transfusion. She was transferred to the Intensive Care Unit (ICU) for further management.

Postoperative laboratory examination revealed a decrease in platelet count (53 000/µL), with stable hemodynamics. She received a minimal dose of dobutamine and morphine. She was extubated 1 day after surgery and moved to the intermediate ward the day after. Serial platelet monitoring was conducted, and the platelet count had increased to 87 000/µL the following day and further increased to 147 000/µL on the third day. The platelet count trend is shown in **Figure 4**. The drainage was reportedly 150 cc/15 hour, or around 5-30 cc/hour.

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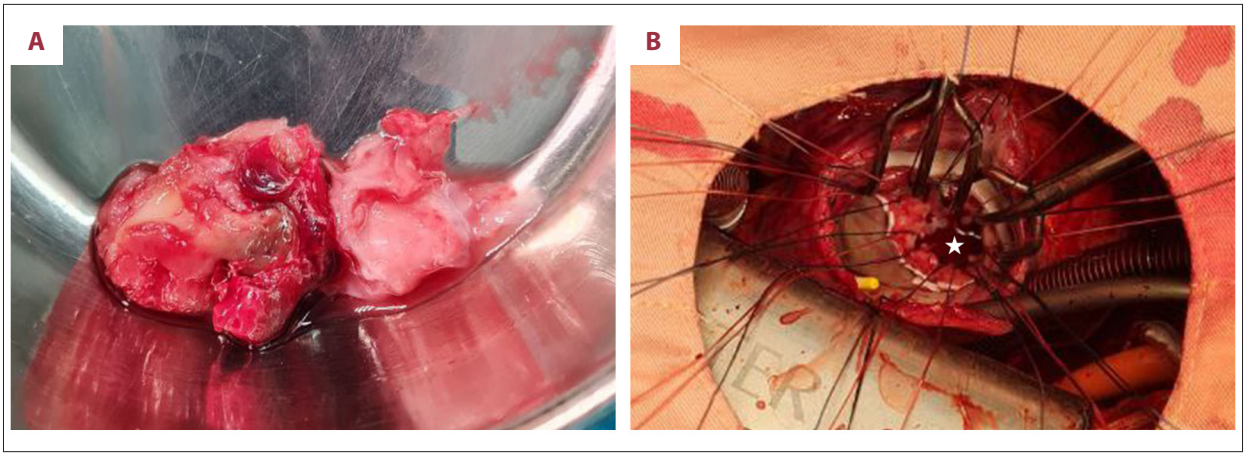

Figure 2. (**A**) Intraoperative documentation: thrombus found in the left atrial. (**B**) Intraoperative documentation before bioprosthetic implantation. White star: MV condition after calcified leaflets and commissure fused were excised.

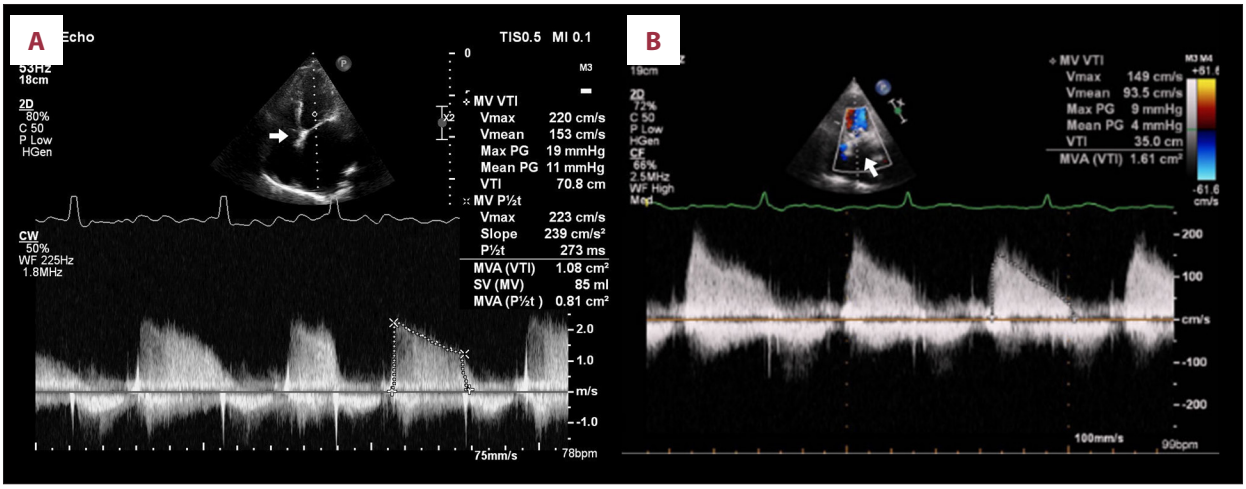

Figure 3. (**A**) Preoperative TTE. The mitral valve appears rheumatic. There is moderate mitral regurgitation and severe mitral stenosis. White arrow: There is a doming of the mitral valve with the restricted opening. (**B**) Postoperative TTE. After MVR with bioprosthetic. The prosthetic valve is well-seated. The prosthetic valve opened well. Doppler evaluation showed normal prosthetic mitral valve gradient, mean MVG 4 mmHg, MVA (VTI): 1.6 cm^2. White arrow: No leakage is seen.

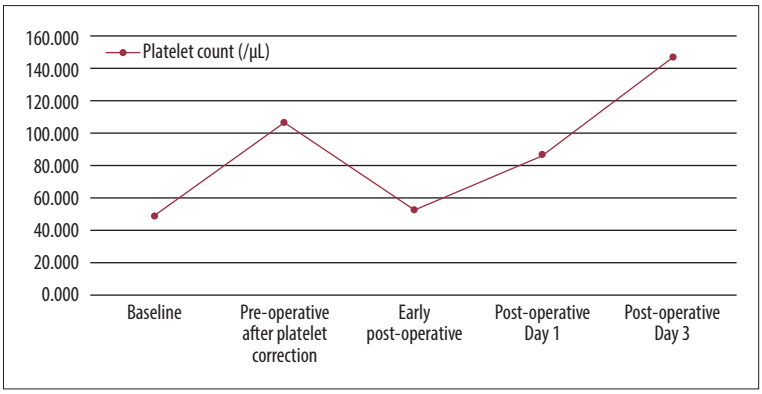

Figure 4. Platelet count trend graph during the hospital stay. Before surgery, laboratory work showed thrombocytopenia (49 000/μL), which was used as the baseline. The platelet count then increased to 107 000/μL after correction. The trend then went down after the patient underwent surgery (53 000/μL). As the treatment for ITP continued, the platelet count continued to increase.

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There were no excessive bleeding or remarkable signs and symptoms upon further monitoring. She was discharged after 8 days of treatment.

Discussion

Immune thrombocytopenia (ITP), formerly known as idiopathic thrombocytopenic purpura, is an acquired autoimmune disorder characterized by an isolated low platelet count of less than 100 000/µL) [1,2]. The main characteristic of ITP is increased peripheral destruction of platelets, with most patients exhibiting anti-platelet membrane glycoproteins antibodies, while the term chronic ITP refers to cases of ITP which last longer than 12 months [3]. Our patient had routinely visited an internist for the past 20 years and had never shown any symptoms of spontaneous bleeding or menstruation abnormalities.

Thrombocytopenia upon hospital admission may be used as a prognostic indicator in RHD patients undergoing valve replacement surgery. According to extensive epidemiological data, thrombocytopenia can also have a direct impact on prognosis. Patients who lost much blood during surgery tend to have negative outcomes. A poor prognosis may be indicated by thrombocytopenia, which may also indicate more severe right-sided heart failure, aberrant hemodynamics (due to hepatic congestion and the ensuing hypersplenism), and abnormal hemodynamics. According to previous research, RV failure is a substantial predictor of cardiovascular morbidity and mortality in patients with heart failure [4].

Thrombocytopenia has been studied in various cardiovascular procedures and is associated with a higher incidence of adverse outcomes, increased mortality rate, and higher health care costs [5,6]. Patients with ITP who undergo cardiac surgery with extracorporeal circulation have an increased risk of postoperative bleeding, postoperative infection, acute kidney injury, and mortality [7-9]. The use of cardiopulmonary bypass (CPB) is also known to decrease platelet count by 25-45% compared to preoperative levels due to inflammatory response to CPB, hemodilution, hypothermia, mechanical damage, and pharmacological treatment (heparin and protamine) [7-12]. Platelet counts tend to decrease 48-72 hours after CPB use, increasing the risk of more decrease due to sepsis, post-transfusion thrombocytopenia, or drug-induced or thrombotic thrombocytopenic purpura [8-11]. Serial platelet monitoring is crucial, especially for patients with a coagulation disorder [7]. In this patient with postoperative serial platelet monitoring, the platelet count was above 50 000/µL, which continuously improved even without platelet transfusion.

Although there is incomplete agreement on the platelet count standard for patients with ITP who will undergo surgery, the existing guidelines suggest that those undergoing major surgery must have a platelet count of at least 50 000/µL and at least 100 000/µL for surgery that has a higher risk of bleeding (brain or heart surgery) [13,14]. In our case, we intended to reach 100 000/µL for safety considerations. The guidelines of ITP management also suggest that some drugs can help reduce the platelet count. The most common and widely used drugs are corticosteroids, which were also used in our patient [15]. Based on the American Society of Hematology (ASH) guidelines for ITP, intravenous immunoglobulin (IVIG) and corticosteroids are used as first-line treatment for ITP in adults who require a rapid increase in platelet count. However, IVIG was not used in our patient because the availability of IVIG in Indonesia is still limited, and the patient has a long history of good response to platelet transfusion and corticosteroids only [16].

The potential risk of platelet transfusion must be considered in aggressive platelet transfusion [17]. The risk of allergic reaction, which is the most common adverse event, must be considered, although it did not happen in our patient [17]. The incidence of left atrial thrombus that occurred in our patient can be considered as an adverse effect of aggressive platelet transfusion, as reports show that thromboembolism is independently linked with platelet transfusion [18,19].

The outcome of this patient was remarkable despite having ITP and undergoing MVR surgery. As we know, physicians have had to deal with patients receiving valve prostheses who had perioperative thrombocytopenia. The shear stress causes the primary mechanism of hemostatic dysfunction through an artificial device, which is accompanied by conflicting mechanisms such as platelet activation, aggregation, and generation of procoagulant microparticles, platelet dysfunction, loss of surface receptors, and bleeding complications. According to the hemodynamic theory, some implant techniques that extend 3 triangles of annular tissue through the implant and into the inflow side of the valve orifice can cause turbulent blood flow, which adds to damage of platelets [12,20]. Therefore, transcatheter valve replacement without CPB and surgical valve replacement with CPB may result in lower platelet counts.

Conclusions

Our case report shows that aggressive preoperative platelet count correction can reduce the risk of mortality and morbidity in patients with ITP who undergo MVR.

Acknowledgement

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

Abbreviations

ITP – immune thrombocytopenia; **MVR** – mitral valve replacement; **TC** – thrombocyte concentrate; **TTE** – transthoracic echocardiography; **MS** – mitral stenosis; **MR** – mitral regurgitation; **RHD** – rheumatic heart disease; **TR** – tricuspid regurgitation; **RV** – right ventricle; **TEE** – transesophageal echocardiography; **RA** – right atrium; **IAS** – intra-atrial septal; **LA** – left atrium; **LAA** – left atrial appendage; **MV** – mitral valve; **ICU** – Intensive Care Unit; **CPB** – cardiopulmonary bypass.

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