

Successful type A aortic dissection repair in the setting of severe immune thrombocytopenia



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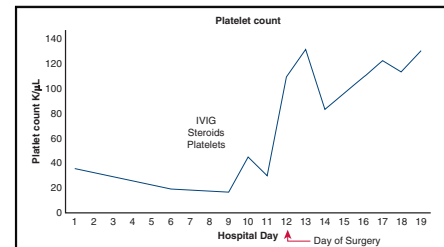
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Management and platelet count of IT patient undergoing type A dissection repair.

CENTRAL MESSAGE

Patients with IT with dangerously low platelet counts can successfully undergo invasive cardiac procedures with aggressive perioperative management.

Video clip is available online.

Platelets play a crucial role in primary hemostasis and control of operative bleeding.¹ Immune thrombocytopenia (IT; previously idiopathic thrombocytopenic purpura or immune thrombocytopenic purpura) is characterized by auto-immune destruction of platelets, bleeding, and surgical complications. IT further increases the risk of perioperative complications during emergent type A aortic dissection (TAAD) repair as it requires hypothermia and circulatory arrest, leading to platelet dysfunction from both IT and cardiopulmonary bypass. We report a patient with severe thrombocytopenia (platelets 17,000/ μ L) who underwent a successful aortic root and hemiarch replacement for TAAD following emergent treatment of IT. Institutional review board approval was waived because patient was deidentified.

CASE DESCRIPTION

A 52-year-old male patient with hypertension and obesity presented with dyspnea. Computed tomography angiography revealed TAAD. Echocardiogram demonstrated severe aortic valve insufficiency. He was initially medically managed due to a platelet count of 17,000/ μ L and a new diagnosis of IT. Laboratory findings showed elevated lactate dehydrogenase and international normalized ratio and low fibrinogen. Peripheral blood smear showed significant schistocytes, reticulocytosis, tear drop cells, and helmet

cells (Video 1). He was treated with methylprednisolone, intravenous immunoglobulin (IVIg), and platelet transfusion on day 9 and 10. Platelets rose to 40,000/ μ L. He required norepinephrine due to hemodynamic compromise from aortic insufficiency and imaging demonstrated pulmonary edema. Institutional review board approval was waived because patient was deidentified.

On day 11, he received 2 units (U) of fibrinogen, 2 U of platelets, a third dose of 30 mg of IVIg, and 12 mg of dexamethasone, and his platelets rose to 110,000/ μ L. On day 12, he underwent aortic root, ascending aortic, and hemiarch replacement. Surgical approach included deep hypothermic circulatory arrest and selective antegrade cerebral perfusion via the right axillary artery. A bio-Bentall valved conduit was created using a size 29 Edwards Magna Ease aortic valve (Edwards Lifesciences) and size 32 Hemashield platinum graft (Maquet Medical Systems). At a temperature of 24 °C, a size 30-mm Hemashield platinum vascular graft was anastomosed in a peninsula-style fashion to the under-surface of the aortic arch. Following reinstatement of body perfusion and during rewarming, the aortic root replacement was performed, and the 2 grafts anastomosed. Circulatory arrest time was 27 minutes, crossclamp time was 192 minutes, and cardiopulmonary bypass time was

SUCCESSFUL TYPE A AORTIC
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THROMBOCYTOPENIA

VIDEO 1. Type A dissection repair with setting of immune thrombocytopenia. Video available at: [https://www.jtcvs.org/article/S2666-2736\(22\)00366-7/fulltext](https://www.jtcvs.org/article/S2666-2736(22)00366-7/fulltext).

246 minutes. Topical hemostats included BioGlue (CryoLife), FloSeal (Baxter), NuKnit (Ethicon), and QuikClot Z-fold gauze (Z-Medica). Intraoperative hematologic testing was guided by hematology consult, and thromboelastography was not recommended. He received 6 U of packed red blood cells, 4 U of platelets, 4 U of fresh-frozen plasma, 1 U of cryoprecipitate and 10 g of aminocaproic acid. He received 2 U of packed red blood cells postoperatively and was discharged on postoperative day 7 with a platelet count of 131,000/ μ L. Computed tomography angiography at 4 months demonstrated no complications. He is now 4 months after surgery without complications.

COMMENT

IT is prevalent in 12.1 per 100,000 adults per year² and is usually a diagnosis of exclusion.^{2,3} While surgery is often offered to a patient with mild/moderate thrombocytopenia ($>70,000/\mu$ L), successful cardiac surgery has rarely been reported with significantly low platelet counts ($<40,000/\mu$ L). Current guidelines are based on expert opinion, without specific platelet count threshold defined. Corticosteroids and IVIG are traditional frontline therapies to

transiently elevate platelet counts. Other therapies include rituximab, azathioprine, splenectomy, thrombopoietin receptor antagonists (romiplostim), and immunosuppressants. Splenectomy is only reserved for patients with steroid-refractory IT,⁴ and romiplostim is currently only on-label for patients with thrombocytopenia as a result of chronic liver disease.⁵

In this patient, steroids and IVIG were sufficient to reach platelet levels over 100,000/ μ L. He received significant transfusions, ie, 9 U of platelets. Platelet age was not tracked. Platelet transfusion is an independent risk factor for complications including nosocomial infections.

This patient survived 12 days of medical management. Considerations in managing patients with thrombocytopenia undergoing cardiac surgery include severity, duration, and etiology of thrombocytopenia. This case demonstrates that with appropriate and aggressive management, it is possible to safely operate on patients with dangerously low platelets. This approach may allow others with severe hematologic abnormalities to undergo cardiac surgery.

In conclusion, a low platelet count due to IT should not automatically exclude cardiac surgery but should be investigated by a multidisciplinary team and managed aggressively. The benefits and risks of preoperative transfusions and advanced treatments should also be considered.

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