Successful Type A aortic dissection repair in the setting of severe Immune Thrombocytopenia.

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Central message: IT patients with dangerously low platelet counts can successfully undergo invasive cardiac procedures with aggressive pre- and peri-operative management.

Introduction:
Platelets play a crucial role in primary hemostasis and control of operative bleeding. IT (previously idiopathic thrombocytopenic purpura or immune thrombocytopenic purpura) is characterized by autoimmune 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 severely thrombocytopenic patient (platelets 17,000/μL) who underwent a successful aortic root and hemiarch replacement for TAAD following emergent treatment of IT. IRB approval was waived.

Case description:
A 52-year-old obese, hypertensive male presented with dyspnea. CT angiography (CTA) revealed TAAD. Echocardiogram demonstrated severe aortic valve insufficiency. He was initially medically managed due to a platelet count of 17,000/μL and a new diagnosis of IT. Laboratory findings showed elevated LDH and INR and low fibrinogen. Peripheral blood smear showed significant schistocytes, reticulocytosis, tear drop cells and helmet cells (video). He was treated with methylprednisolone, IVIG and platelet transfusion on day 9 and 10. Platelets rose to 40,000/μL. He required norepinephrine due to hemodynamic compromise from aortic insufficiency and imaging demonstrated pulmonary edema. IRB approval waived because patient was deidentified.

On day 11, he received 2 units (U) of fibrinogen, 2U platelets, a third dose of 30mg IVIG, and 12mg dexamethasone and his platelets rose to 110,000/μL. On day 12, he underwent aortic root, ascending aortic and hemi-arch replacement. Surgical approach included deep hypothermic circulatory arrest and selective antegrade cerebral perfusion via the right axillary artery. A biobentall valved conduit was created using a size 29 Edwards Magna ease aortic valve (Edwards Lifesciences, Irvine, CA) and size 32 Hemashield platinum graft (Maquet Medical Systems, Wayne, NJ). At a temperature of 24°C, a size 30-mm Hemashield platinum vascular graft was anastomosed in a peninsula-style fashion to the undersurface of the aortic arch. Following re-institution of body perfusion and during rewarming, the aortic root replacement was performed, and the two grafts anastomosed. Circulatory arrest time was 27 minutes, cross clamp time was 192 minutes and cardiopulmonary bypass (CPB) time was 246 minutes. Topical hemostats included Bioglue (Cryolife, Kennsaw, GA), FloSeal (Baxter, Wyayrd, CA), NuKnit (Ethicon, Cincinnati, OH), and QuikClot Z-fold gauze (Z-Medica, Wallingford, CT). Intra-operative hematological testing was guided by hematology consult and Thromboelastography was not recommended. He received 6U packed red blood cells (PRBCs), 4U platelets, 4U fresh frozen plasma, 1U cryoprecipitate and aminocaproic acid. He received 2U PRBCs post-operatively and was discharged on post-operative day 7 with a platelet count of 131,000/μL. CTA 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 year\(^2\) and is usually a diagnosis of exclusion\(^2,3\). While surgery is often offered to a patient with mild/moderate thrombocytopenia (>70,000/μL), successful cardiac surgery has rarely been reported with significantly low platelet counts (<40,000/μL). 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 steroid refractory IT patients\(^4\) 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/μL. He received significant transfusions, 9 units of platelets. Platelet transfusion is an independent risk factor for complications including nosocomial infections.

This patient survived 12 days of medical management. Considerations in managing thrombocytopenic patients 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 investigated by a multidisciplinary team and managed aggressively. The benefits and risks of preoperative transfusions and advanced treatments should also be considered.

References:
Central Picture Legend: Management and platelet count of IT patient undergoing type A dissection repair.

Video Legend: Type A Dissection Repair with setting of IT.
Platelet count

IVIG
Steroids
Platelets

Hospital Day

Day of Surgery
SUCCESSFUL TYPE A AORTIC DISSECTION REPAIR IN THE SETTING OF IMMUNE THROMBOCYTOPENIA