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# Splenectomy in a patient with chronic idiopathic thrombocytopenic purpura with critical thrombocytopenia, an anesthetic challenge - A case report

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***Abstract*---**Immune thrombocytopenic purpura, also termed as idiopathic thrombocytopenic purpura is an acquired disorder in which there is immune mediated destruction of platelets. Here we describe the perioperative management of a case of chronic ITP undergoing splenectomy. Anesthetic management of the patient with ITP includes general anesthesia, avoiding NSAIDs and platelet lowering drugs, transfusing platelets, avoiding airway trauma and watching for hemorrhagic complications.

***Keywords*---**immune thrombocytopenic purpura, critical thrombocytopenia, anesthetic challenge.

## Introduction

Immune thrombocytopenic purpura, also termed as idiopathic thrombocytopenic purpura is an acquired disorder in which there is immune mediated destruction of platelets and possibly inhibition of platelet release from the megakaryocyte[1] .In children, it is usually an acute disease, with self limited course. However, in adults it is a more chronic disease. It is termed as secondary, if it is associated with an underlying autoimmune disorder particularly SLE and infections like HIV, Hepatitis C.[1]

The first-line therapy for ITP is oral corticosteroids. Splenectomy is generally considered a second-line therapy in patients who are refractory to steroids and in those who relapse after an initial response to medical therapy.[2]Anesthetic management in patients with ITP is quite challenging because of increase risk of perioperative complications.

**Case Report**

We report a case of 22 years old unmarried female who presented to medical outpatient department with complaints of gum bleeding, menorrhagia , nasal bleed in February 2020. On examination, there were petechia present, facial rash and oral ulcers. Routine investigations were done , which showed Platelet counts 5000/$mm^3$, Hemoglobin(Hb) 11.3 g/dl ,Total Leucocyte Counts(TLC) 6.8/$mm^3$ ,
Liver Function Test, Kidney Function Test, electrolytes, coagulogram were normal. ANA -positive , Anti -ds DNA -negative , ICT-negative , DCT- positive , Hepatits serology -negative ; (Anti histone , Anti centromere ,Scl 70 , Jo-1 ,CRP ,RF, Anti CCP)- negative. Bone marrow aspiration showed - features suggestive of dual deficiency anemia with no neoplastic pathology.

PBF showed - RBCs: with decreased density with normocytic normochromic with few microcytic hypochromic and occasional macrocytes seen. Platelets: marked thrombocytopenia on peripheral blood film approximately <5000/$mm^3$. WBCs: N 52/ L38/ M07/E01.
USG abdomen - showed normal study

She received pulse steroid therapy during her stay in hospital and her platelet counts improved to 31000/$mm^3$, after which she was discharged and was put on haematology follow up. During her course of treatment she also received, mycophenolate mofetil (Immunomodulator), dapsone, eltrombopag(Thrombopoetin receptor agonist) apart from steroids.

In September 2021, she was admitted again with similar complaints, with platelets of 17000/$mm^3$. She was given Iv Ig, however, there was no improvement in her platelet count and was finally planned for laparoscopic splenectomy.She continued to have her routine medicines as advised by hematologist(eltrombopag , steroids , tranexemic acid). One week prior to surgery her platelet counts fell to lowest of 1000/$mm^3$, with no signs of spontaneous bleed from any orifice. For this she received one unit of SDAP .A combined approach in collaboration with surgical team, clinical hematologist and anesthesia team was done perioperatively.On the day of surgery her platelet counts were 19000/$mm^3$. 160 ml of SDAP was given 2 hours prior to surgery. She was given iv antibiotics half an hour prior to surgery. After receiving her in OR, multi channel monitor was connected. . Her preoperative heart rate was 90 bpm, Blood Pressure 128/92 mmHg, $SPO_2$ 97%. 20G iv canula was in place. Another 18 G iv canula was secured. She was given 250 ml of SDAP before induction. Premedication was done with inj pantoprazole 40 mg iv. Preoxygenation was done with 100% oxygen for 3 minutes. Patient was induced with Inj fentanyl 60mcg iv, inj propofol 120mg iv, inj atracurium 30 mg iv, sevoflurane 2%. Direct laryngoscopy was done smoothly and airway was secured with cuffed ETT 7.0mm ID atraumatically. Maintenance of anesthesia was continued with 40% oxygen , 60% nitrous oxide , isoflurane

with MAC around 1.0 and intermittent boluses of inj atracurium 5mg. Patient was also given inj dexamethasone mg iv ,inj hydrocortisone 100 mg iv, inj methylprednisolone 1 g iv , inj tranexemic acid 1 g iv. 16 FG nasogastric tube was placed smoothly in first attempt after preparing nostril with xylometazoline nasal drop and adequate lidocaine 2% jelly. However, patient had nasal bleed soon after it was placed and secured. Nasal pack was placed and topically botroclot (haemcoagulase enzyme) nasal drops were instilled after which the bleeding was controlled. She was catheterized with foley's catheter 16F atraumatically. Prior to surgical incision, her Complete Blood Count was sent which showed Hb8.5g/dl , TLC 9900/ $mm^3$ , platelets 43000/$mm^3$. For analgesia, she received additional 40mcg of inj fentanyl iv, inj 100mg of tramadol iv, 1g inj paracetamol iv. NSAIDs were avoided. No intamuscular injections were given and regional blocks were avoided. She also received intraoperatively 160 ml of SDAP, 3 units of RDP as bleeding continued from surgical site .Surgery continued for 4 hours and vitals were maintained within normal limits. An estimated 200-300 ml of blood loss was calculated. Total of 2.5 L of crystalloids were given and one unit of PRBCs. Urine output of 800 ml was recorded and there were no signs of hematuria. After ligation of splenic vessels, repeat CBC showed platelets of 36000/ $mm^3$, Hb 7.1g/dl. Throughout the procedure patient remained hemodynamically stable. At the end of surgery, patient was given antiemetic inj ondensetron 6mg iv and was reversed once she started breathing spontaneously with inj Neostigmine @ 60mcg/kg +inj Glycopyrolate @10 mcg/kg. Extubation was smooth and uneventful. Patient was conscious and was obeying commands properly. Due to lack of availability of High Dependency Unit in our hospital, she was shifted to ICU for further monitoring. Post operative CBC showed improvement in her platelet counts from 43000/$mm^3$ to 71000/$mm^3$ on zero Post Operative Day and 86000/$mm^3$ on 1$^{st}$ Post Operative Day.

**Discussion**

The incidence of ITP is estimated to be 2 to 5 per 100 000 persons in the general population[3,4,5]. Large randomized trials on the management of ITP are lacking, resulting in significant controversy and variation in practice. During ITP, it has been observed that brief, spontaneous remissions can occur frequently in children. On the other hand, adult patients rather display a more chronic form of ITP that correlates with significant clinical presentations including bleeding disorders, hemorrhages in skin or mucous membranes, namely purpura, petechiae and rarely intracranial manifestations of the disease[6,7].Treatment strategies for ITP are mostly prescribed on the basis of clinical symptoms of the patients with a focus on reducing the risk of severe bleeding, and they do not essentially include the boosting of platelet numbers.In adults with newly diagnosed ITP and a platelet count of >/=30*$10^9$ /L who are asymptomatic or have minor mucocutaneous bleeding, the ASH (American society of haematology)guideline panel recommends against corticosteroids and in favor of management with observation and adults with newly diagnosed ITP and a platelet count of <30 * $10^9$ /L who are asymptomatic or have minor mucocutaneous bleeding, the American Society of Hematology (ASH) guideline panel suggests corticosteroids rather than management with observation. Those with ITP lasting >3 months who are corticosteroid-dependent or have no response to corticosteroids, the ASH guideline panel suggests either splenectomy or a TPO-

RA[8] .With regards to type of surgery , whether laparoscopic or open splenectomy; laparoscopic splenectomy is associated with shorter hospital stay , less blood loss and blood transfusion during surgery than open splenectomy[9,10]. However, hematological outcomes are not different to conventional splenectomy[11]

These patients pose risk of life threatening hemorrhage and major anesthetic and surgical challenge to achieve hemostasis.The bleeding risk increases with age and when the platelet count is less than 20-30 x $10^9$/L. The mean platelet volume in ITP increases as the platelet count decreases and the larger platelets are assumed to be younger and more reactive. Hence the bleeding time (BT) of patients with thrombocytopenia is shorter than expected for the degree of thrombocytopenia ; and severity of bleeding in ITP is less than that seen in comparable thrombocytopenia in bone marrow failure[12,13].

Important anesthetic considerations for patients with ITP undergoing splenectomy are to build up platelets prior to surgery using either SDAP or RDP. One unit of Random Donor Platelet are expected to raise platelet count by 3000-5000/$mm^3$. While one unit of SDAP(Single Donor Apheresis Platelets) increases by 40000-60000/ $mm^3$.[14] Thromboelastography can be a very helpful tool intraoperatively in such scenarios Drugs that cause platelet dysfunction like NSAIDS need to be avoided. General anesthesia is usually given to such patients, however, airway manipulation needs to be minimum and gentle. Because of risk of spinal hematoma and the associated morbidity with it , neuraxial anesthesia needs to be avoided in patients with thrombocytopenia . Patients planned for splenectomy should received vaccination against encapsulated microorganisms like pneumococcus and meningococcus. However,despite pneumococcal vaccination prior to splenectomy; fatal fulminant sepsis is an omnipresent possibility . Such patients also need close follow up in post operative period for hemorrhagic complications. We need to do serial blood tests to see improvement in platelet counts .[15]Approximately 80% of the patients respond to splenectomy, among which approximately 66% experience longterm remission without further therapy .[16] As the time since splenectomy increases, the rate of excellent response decreases[17].Despite recent developments, the expected increase in the success rate of treatments has not been achieved yet. A substantial number of patients either do not respond at all or respond only transiently to many treatment interventions.As yet, there is no consistently effective method to predict an individual ITP patient's response to splenectomy.

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