Anesthetic management of laparoscopic splenectomy in a case of Evans Syndrome with systemic lupus erythematos

Madam,

Evans Syndrome is a rare autoimmune disorder and by definition is the combination of both concurrent and sequential development of idiopathic thrombocytopenic purpura (ITP) and Coombs positive autoimmune hemolytic anemia (AIHA) in the absence of underlying disorder. Although the true incidence is not known, it is estimated to effect 3.7-5% of patients with ITP or AIHA at the onset. The dysregulation of the immune system produces several autoantibodies against red blood cells and platelets leading to early destruction by complement and reticuloendothelial systems. The spleen has been proposed as either a site of destruction or as a source of autoantibodies production. The presence of associated systemic lupus erythematos (SLE), warrants a more aggressive line of management than either condition presenting alone with poorer prognosis. We present case of Evans syndrome with ongoing bleeding diathesis, anticipated anesthetic risks and challenges scheduled for elective laparoscopic splenectomy in the presence of depleted platelet count.

A 26-year-old female patient presented with bleeding gums, menorrhagia and petechiae for the last 1-month, and 3 years back she had similar complaints along with jaundice. She was diagnosed to have “steroid-resistant Evans syndrome” associated with SLE. She had remission with two doses of rituximab. The present medication consisted of oral prednisolone 30 mg, hydroxychloroquine 200 mg, injection insulin and calcium supplements. General examination revealed petechiae over trunk and limbs. Her investigations are shown in Table 1. Electrocardiogram, two-dimensional echocardiogram and chest X-ray was normal. Ultrasonography of the abdomen revealed mild splenomegaly and distended gallbladder. She has a short neck with Mallampati Grade II. Vital parameters were normal except mild tachycardia. She was vaccinated for pneumococcal, meningococcal and haemophilus infections. Premedication consisted of oral pantoprazole; alprazolam and rest of the medications were continued except insulin.

On the day of surgery baseline, blood glucose was 65 mg/dL and corrected to 156 mg/dL with 25% dextrose. A large bore 16G intravenous (IV) cannula was secured; 1 unit single donor platelets (SDP) and hydrocortisone 25 mg IV was given prior to the surgery. Anesthesia was induced, and oral endotracheal tube was placed with minimal manipulation. Crepe bandages were applied to the lower limbs. Intraoperative analgesia was with IV fentanyl 80 µg bolus followed by 20 µg slow IV as required. Another unit of SDP and 3 units of platelet rich plasma and 2 units of packed red cells were given to counter the blood loss of approximately 800 mL. The procedure lasted for 5 ½ h with stable hemodynamics, and the urine output was 400 mL. Postoperative course was uneventful; IV hydrocortisone 25 mg was repeated every 6th hourly for 24 h. Platelet count was 70,000 mm$^3$, and there was no bleeding. She was discharged home on 10th postoperative day. Follow-up after 1-month was unremarkable.

Anesthetic management of laparoscopic splenectomy in a case of Evans Syndrome with systemic lupus erythematos

Madam,

Evans Syndrome is a rare autoimmune disorder and by definition is the combination of both concurrent and sequential development of idiopathic thrombocytopenic purpura (ITP) and Coombs positive autoimmune hemolytic anemia (AIHA) in the absence of underlying disorder. Although the true incidence is not known, it is estimated to effect 3.7-5% of patients with ITP or AIHA at the onset. The dysregulation of the immune system produces several autoantibodies against red blood cells and platelets leading to early destruction by complement and reticuloendothelial systems, The spleen has been proposed as either a site of destruction or as a source of autoantibodies production. The presence of associated systemic lupus erythematos (SLE), warrants a more aggressive line of management than either condition presenting alone with poorer prognosis. We present case of Evans syndrome with ongoing bleeding diathesis, anticipated anesthetic risks and challenges scheduled for elective laparoscopic splenectomy in the presence of depleted platelet count.

A 26-year-old female patient presented with bleeding gums, menorrhagia and petechiae for the last 1-month, and 3 years back she had similar complaints along with jaundice. She was diagnosed to have “steroid-resistant Evans syndrome” associated with SLE. She had remission with two doses of rituximab. The present medication consisted of oral prednisolone 30 mg, hydroxychloroquine 200 mg, injection insulin and calcium supplements. General examination revealed petechiae over trunk and limbs. Her investigations are shown in Table 1. Electrocardiogram, two-dimensional echocardiogram and chest X-ray was normal. Ultrasonography of the abdomen revealed mild splenomegaly and distended gallbladder. She has a short neck with Mallampati Grade II. Vital parameters were normal except mild tachycardia. She was vaccinated for pneumococcal, meningococcal and haemophilus infections. Premedication consisted of oral pantoprazole; alprazolam and rest of the medications were continued except insulin.

On the day of surgery baseline, blood glucose was 65 mg/dL and corrected to 156 mg/dL with 25% dextrose. A large bore 16G intravenous (IV) cannula was secured; 1 unit single donor platelets (SDP) and hydrocortisone 25 mg IV was given prior to the surgery. Anesthesia was induced, and oral endotracheal tube was placed with minimal manipulation. Crepe bandages were applied to the lower limbs. Intraoperative analgesia was with IV fentanyl 80 µg bolus followed by 20 µg slow IV as required. Another unit of SDP and 3 units of platelet rich plasma and 2 units of packed red cells were given to counter the blood loss of approximately 800 mL. The procedure lasted for 5 ½ h with stable hemodynamics, and the urine output was 400 mL. Postoperative course was uneventful; IV hydrocortisone 25 mg was repeated every 6th hourly for 24 h. Platelet count was 70,000 mm$^3$, and there was no bleeding. She was discharged home on 10th postoperative day. Follow-up after 1-month was unremarkable.

Anesthetic management includes a thorough preoperative assessment, perioperative continuation of steroid/immunosuppression and an additional dose of steroid for possible suppression of hypothalamo-pituitary adrenal axis and evaluation of systemic organ involvement. Airway assessment may reveal unanticipated difficult airway,

Table 1: Laboratory investigations

| Parameter (units) | Result | Reference range |
|------------------|--------|-----------------|
| Fasting sugar (mg/dL) | 139 | 90-110 |
| Blood urea (mg/dL) | 22 | 10-40 |
| Serum creatinine | 0.9 | 0.7-1.2 |
| Hemoglobin (g/dL) | 10 | 12-15 |
| Total leucocyte count (mm$^3$) | 7700 | 4000-10,000 |
| Platelet count (mm$^3$) | 50,000 | 1.4 |
| Total proteins (g/dL) | 5.3 | 6-7.5 |
| Albumin (g/dL) | 3.5 | 3-5 |
| Total bilirubin (mg/dL) | 5 | 0.1-0.8 |
| Conjugated bilirubin | 0.3 | |
| Prothrombin time (sec) | 13.6 | 10-13 |
| APTT (sec) | 27.9 | 22-32 INR 1.2 |
| SGOT (U/L) | 42 | Up to 40 |
| SGPT (U/L) | 22 | 5-45 |
| LDH (IU/L) | 661 | 0-480 |
| Anti-dsDNA (IU/mL) | 15 | 50 |
| aCl Ab IgG (GPL/mL) | 72 | <40 |
| aCl Ab IgM (GPL/mL) | 6 | <40 |
| ANA ++ homogenous |
| Direct Coombs test | +++
| Indirect Coombs test | + |
| Lupus anticoagulant | Negative |
| Antiphospholipid antibody | Negative |

APTT = Activated partial thromboplastin time, SGOT = Serum glutamic-oxaloacetic transaminase, SGPT = Serum glutamic pyruvic transaminase, LDH = Lactate dehydrogenase, aCl = Anticardiolipin, Ab = Antibody, IgG = Immunoglobulin G, IgM = Immunoglobulin M, ANA = Antinuclear antibody, Anti-dsDNA = Anti-double-stranded DNA, INR = International normalized ratio
subglottic/laryngeal edema. Avoid trauma and bleeding during airway management. Strict asepsis must be maintained as these patients are at intrinsic susceptibility and also immunosuppressants. A careful preanesthetic evaluation, a definitive anesthetic strategy with special considerations to risk of airway bleed, thrombocytopenia, systemic organ involvement and perioperative steroid replacement is mandatory.

Nirmala Jonnavithula, Sai Lakshman Pasupuleti, Venumadhav Thumma, Gopinath Ramachandran

Departments of Anesthesiology and Intensive care, Surgical gastroenterology, Nizam’s Institute of Medical Sciences, Panjagutta, Hyderabad, Telangana, India

Address for correspondence: Dr. Nirmala Jonnavithula, Departments of Anesthesiology and Intensive care, Nizam’s Institute of Medical Sciences, Panjagutta, Hyderabad - 500 082, Telangana, India.
E-mail: njonnavithula@gmail.com

References

1. Evans RS, Takahashi K, Duane RT, Payne R, Liu C. Primary thrombocytopenic purpura and acquired hemolytic anemia; evidence for a common etiology. AMA Arch Intern Med 1951;87:48-65.
2. Norton A, Roberts I. Management of Evans syndrome. Br J Haematol 2006;132:125-37.
3. Wang W, Hershon H, Pai CH, Presbury G, Wilimas J. Immunoregulatory abnormalities in Evans syndrome. Am J Hematol 1983;15:381-90.
4. Savasan S, Warrier I, Ravindranath Y. The spectrum of Evans' syndrome. Arch Dis Child 1997;77:245-8.
5. Kang I, Park SH. Infectious complications in SLE after immunosuppressive therapies. Curr Opin Rheumatol 2003;15:528-34.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Psoas compartment block for intraoperative anesthesia for fracture neck of femur: Case report

Madam,

Delivering anesthesia to high-risk patients who have sustained a fractured neck of femur (NOF) can be challenging. The decision is largely dichotomous with either a general anesthetic (GA) and/or a regional technique which commonly includes a subarachnoid block (SAB) or a combination of a sciatic and lumbar plexus (LP) block. [1,2] LP blocks in singularity are not well-described in the literature.[3] We report an LP block as the sole anesthetic technique in a 66-year-old female. who suffered from dementia, and sustained a left NOF fracture following a fall. She had a significant co-morbid history for ischemic heart disease and myocardial infarction, type 2 diabetes, recent PE and transient ischemic attacks, on clopidogrel. Routine investigations were unremarkable, and she remained hemodynamically stable. Antiplatelet agents contraindicated a SAB, and we hoped to avoid potential cognitive decline and hemodynamic insult associated with a GA, so third party consent was obtained for an LP block with conversion to GA if unsuccessful.

With intravenous access, standard monitoring, nasal oxygen, and midazolam 1.5 mg, she was positioned in the right lateral position. Landmarks were identified, and both an intercrestal line and parasagittal line were marked. At their intersection, following local anesthetic (LA), a 100 mm Stimuplex Insulated Needle (SonoplexStim Cannula 21G Pajunk, Geisingen, Germany) was inserted with a nerve stimulator. Using a loss of resistance technique, the psoas compartment was reached on the second pass and a quadriceps contraction elicited. While maintaining a motor response with <0.4 mA and negative aspiration, a test dose of 2 ml of 1% ropivacaine (Ropivacaine Sandoz, NSW, Australia) was injected. The twitch disappeared, and the patient was able to move her toes....