Section: Anaesthesia

Case Report

ANAESTHETIC MANAGEMENT OF A PATIENT WITH EVANS SYNDROME FOR EMERGENCY PERIANAL ABSCESS DRAINAGE: A CASE REPORT

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Abstract

Evans syndrome was first described by Robert Evan in 1951.It is a very rare autoimmune disorder and defined as the combination of idiopathic thrombocytopenic purpura (ITP) and Coombs positive autoimmune hemolytic anemia (AIHA). Signs and symptoms are variable. Pre anaesthetic evaluation should focus on definitive anaesthetic strategy, considerations to systemic involvement, thrombocytopenia leading to risk of airway bleed and other haemorrhaging complications and management. We present anaesthetic management of a 23 year old patient with Evans syndrome (ES) along with Systemic Lupus Erythematosis (SLE) posted for emergency perianal abscess drainage.

INTRODUCTION

Evans syndrome is a rare autoimmune disorder first described by Robert Evans in 1951. It is defined as combination of both concurrent and sequential development of idiopathic thrombocytopenia purpura (ITP) and coomb's positive autoimmune hemolytic anemia in the absence of underlying disorder.[1] Due to dysregulated immune response, autoantibodies are produced against red blood cells and platelets. Spleen has been proposed as a site of production or destruction of such antibodies.[2,3] Importantly, ES is associated to another diseases in 27-50% of cases, most importantly haematological malignancies and systemic lupus erythematosus in adults.[4,5] We hereby present a case report of 23 years old female and a known case of Evans syndrome with systemic lupus erythematosis posted for emergency incision and drainage of perianal abscess under general anaesthesia.

CASE REPORT

A 23 year old female presented with chief complaints of pain and swelling in perianal region since five days associated with fever. She was diagnosed as a case of perianal abscess and posted for emergency incision and drainage. Patient was a known case of Evans syndrome and was on T. Wysolone and mycophenolate mofetil since one year. Patient had history of easy bruising, bleeding from gums and menorrhagia which raised suspicion of underlying coagulopathy which eventually lead to diagnosis of Evans syndrome with SLE. She was on steroid and immunosuppressive therapy. On general examination patient was febrile. Investigations were Hemoglobin-10.8 gm%, Total leucocyte count-21000/cumm, differential leucocyte count-P80/L18/E1/B1, Platelet count-1.2lac/cumm, PT/INR-17/1.2. Blood urea- 23, Serum creatinine-0.6, SGOT/SGPT/ALP-28/65/80.Ultrasonography of local site was suggestive of 5cm x 3.8cm of collection in left perianal region with surrounding inflammatory changes. We planned neuraxial anaesthesia for the patient as coagulation profile was normal but due to refusal of patient for spinal anaesthesia, general anaesthesia was planned after proper consent of patient. A large bore 16G intravenous cannula was secured. Monitors, noninvasive blood pressure, electrocardiogram, pulse oximeter and end tidal carbon dioxide were connected. Baseline parameters were noted. Inj. hydrocortisone 50mg IV was given prior to surgery. Patient was preoxygenated with 100% O2 for 3 min. Patient was induced with inj. fentanyl 100 mcg iv and inj. propofol 120 mg iv. Inj atracurium 30mg iv was given prior to intubation and patient was intubated gently with oral endotracheal tube of size 7mm internal diameter. Anaesthesia was maintained with sevoflurane. The goal of anaesthesia was to maintain the vital parameters. There was no excessive bleeding and patient remained stable throughout the procedure. Patient was extubated after full neuromuscular recovery. Post operative period was uneventful and patient was shifted to ward after monitoring in post anaesthetic care unit.

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DISCUSSION

Evans syndrome is a very rare autoimmune condition reflecting a major breakdown of immune self tolerance. The etiology of Evans syndrome is unknown and is defined as idiopathic. Signs and symptoms of Evans syndrome are variable and depend on the type of blood cell lines that are involved. Anaesthetic management includes a thorough preoperative assessment and perioperative care. Pre anaesthetic evaluation should focus on definitive anaesthetic strategy, considerations to systemic involvement, thrombocytopenia leading to risk of airway bleed and other haemorrhaging complications. Laryngoscopy should be done gently to avoid trauma and bleeding during intubation.[6] Strict asepsis must be maintained as these patients are at intrinsic susceptibility of infection due to use of immunosuppressants.[7] Perioperatively steroids should be considered as they reduce destruction of platelets and red blood cells by reducing sequesteration. Moreover patients on long term steroid therapy might be at risk of developing acute adrenal crisis hence it might necessitate steroid coverage before surgery. Anaesthetic management of such patient requires cautious use of regional anaesthesia. Regional anaesthesia should be avoided in patients presenting with thrombocytopenia. Choice of anaesthesia should depend on coagulation profile, nature of surgery, general condition of patient and willingness of the patient for the particular anaesthetic procedure. Blood product should be available during the procedure. Use of hemodilutional autologous transfusion for the patient with Evans syndrome is beneficial to minimize immune hemolytic process.[8]

CONCLUSION

Anaesthetic management of a patient with Evans syndromes should include meticulous preoperative anaesthetic checkup, evaluation of associated comorbidities, perioperative concerns and post anaesthetic care. Choice of anaesthesia should be determined by general condition of patient, nature of surgery, coagulation profile and willingness of patient for the specific anaesthetic procedure. Special concern should be laid on possibility of hemorrhagic complications, systemic involvement, use of steroids and autologous blood transfusion.

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