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ANESTHETIC MANAGEMENT OF A PATIENT WITH EVANS SYNDROME FOR ELECTIVE OPEN SPLENECTOMY: A CASE REPORT

Anaesthesiology

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ABSTRACT

Background: Patients with Evan syndrome are a challenge to anesthesiologists due tothe risk of thrombocytopenia,bleeding during airway management and associated risk of haemmoraghic complications.

Case presentation: We present anesthetic management of a 50-year-old Indian lady with secondary Evans syndrome for elective open splenectomy under General Anaesthesia.

Conclusion: patient with Evan syndrome can be managed with meticulouspreanesthetic evaluation, a definitive anesthetic managment with special considerations to risk of airway bleed, thrombocytopenia, systemic organ involvement and perioperative steroid replacement.

KEYWORDS

INTRODUCTION

Evans Syndrome is a rare autoimmune disorder, defined as 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[1].Although the true incidence is not known,it is estimated to affect 3.7-5% of patients with ITP or AIHA at the onset[2]. The dysregulation of immune system produces several auto antibodies against red blood cells and platelets leading to early destruction by compliment and reticuloendothelial system[3].The spleen has been proposed as either a site of destruction or a source of autoantibodies production[1]. The presence of associated autoimmune disorder such as systemic lupus erythematosis (SLE) warrants a more aggressive line of management with poor prognosis.we present a case of Evans syndrome with ongoing bleeding diathesis,anticipated anesthetic risks and challenges scheduled for elective open splenectomy under General Anaesthesia in the presence of depleted platelet count.

CASE PRESENTATION

A 50-year-old Indian lady presented with complaints of bleeding gums, blood tinged expectoration for last 4-5 months and reddish blue patchy skin discoloration since last 5 years. HOPI revealed that she was apparently well 6 years back when she noticed easy bruising of skin after minor trauma for which she took medical advice from different polyclinics and private hospitals. She was diagnosed 2yrears back with Secondary Evan syndrome with Systemic Lupus Erythematosis. Initially she was started on oral Prednisolone 60mg , calcium supplements and responded well to it but few months later due to relapse of symptoms she was rediagnosed as Steroid Resistant Secondary Evan syndrome and started on Rituximab. During course of management she was diagnosed with other comorbidities such as Diabetes mellitus type 2 and Hypothyroidism for which regular insulin, insulin Lente and Tablet Levothyroxine was started respectively.

On general physical examination – petechial patches over trunk and limbs (fig.1,2).Afebrile, pulse rate 72 per min regular,Respiratoryrate 18 breaths per minute and blood pressure 120/70 mmHg.systemic examination suggestive of petechial patch over right paraumblical region on inspection and splenomegaly as palpation finding.

Preoperative laboratoryevaluation reports are given in Table 1.

Patient's electrocardiogram and chest X ray was WNL (Fig 3).

USG ABDOMEN suggestive of mild splenomegaly ,Grade 1 fatty changes in liver.

BONE MARROW BIOPSY suggestive of erythroid hyperplasia, megakaryocytic thrombocytopenia.

DXA SCAN suggestive of osteopenia.

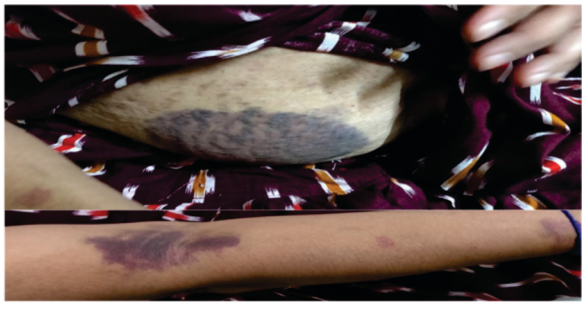

Fig 1,2 :- showing petechial patches over trunk and upper limb

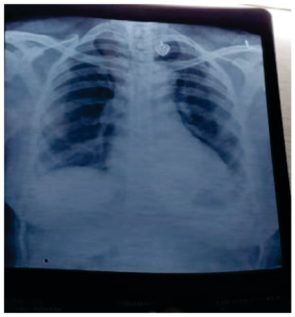

Figure 3 showing chest X-Ray

Table 1 showing preoperative blood investigations.

Haemoglobin	11.5gm%
Total leukocyte count	5300
Differential leukocyte count	P85,L12,M2,E0,B01
Platelet count	10000
PT/PTTK/INR	10.1/33/0.74
RBS	111 mg/dl

Fasting	90
Post prandial	106
HbA1c	12.7
Blood urea	37 mg/dl
Serum creatinine	0.8 mg/dl
S-Na+/k+	136/3.8 mmol/lt
s.bilirubin (Total)	0.4 mg/dl
AST/ALT	21/17 U/L
T.Protein	5.5 g/dl
S.Albumin/Globulins	
Urine routine microscopy	Glucose +++
Thyroid profile	Within normal limits

DISCUSSION

Ourcase had following concerns/issues; Thrombocytopenia, immunosuppression,poor glycemic control,risk of bleeding during airway management and risk of hemorrhagic complications.Decision of elective open splenectomy under general anaesthesia was taken after discussion with multidisciplinary team regarding the risk of bleeding while securing airway, thrombocytopenia, immunosuppression,poor glycemic control, risk of excessive blood loss.ouranaesthesia management included institution of general anaesthesia along with aim to transfuse platelets preferably single donor platelets to prevent allo immunization, intravenous immune globulins or steroids to cut down platelets destruction,monitoring for hemorrhagic complications, abstinence from use of NSAID's or other platelet lowering drugs, avoidance of airway trauma,nasal intubation,intramuscular injections. We ensured patient had taken course of pneumococcal vaccine.

During perioperative management on the day of surgery after shifting in operation theatre all standard monitors were attached and 02 large bore intravenous cannulae were secured. One unit SDP and injection hydrocortisone 25 mg intravenous given prior to surgery. After preoxygenationanaesthesia was induced with propofol and atracurium followed by atraumatic intubation with direct laryngoscopy using 7.5 mm cuffed endotracheal tube. Anaesthesia was maintained with nitrous,oxygen,sevoflurane and atracurium boluses. intraopanalgesia was achieved with intravenous fentanyl. Another unit of SDP and 02 units of PRBC were given to counter blood loss of approximate 1000 ml. procedure lasted for about 3hours with stable hemodynamics and urine output of 600 ml.patient extubated after full neuromuscular recovery. Post operative period was uneventful.patient shifted to respective ward.post operative platelet count was 70000 There was no bleeding and patient was discharged home on 10thpost operative day and was under follow up with hematology department.

CONCLUSION

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 di cult airway, 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[5]. 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.

COMPETING INTEREST

The authors declare that they have no competing interests.

AUTHORS' CONTRIBUTIONS

All of the authors were involved in the management of thecase and finalizing the article. All of the authors wereinvolved in the process of editing, correcting, and finalizingthe manuscript. All authors have read and approvedthe final manuscript.

CONSENT

The patient and the patient's relatives are pleased to givewritten consent for publishing this particular casescenario.

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