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Original Research Paper

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ANAESTHETIC MANAGEMENT IN A PARTURIENT WITH EVAN'S SYNDROME AND HYPOTHYROIDISM FOR CESAREAN SECTION.

Dr. Pooja Chaudhari — Junior resident, Department of Anaesthesia, Seth GS Medical College and KEM Hospital, Parel, Mumbai

Dr. Priti Devalkar — Associate Professor, Department of Anaesthesia, Seth GS Medical College and KEM Hospital, Parel, Mumbai

ABSTRACT Evans syndrome is a rare autoimmune condition with a chronic relapsing course. Patients with Evans syndrome are a challenge to the anaesthesiologist due to the risk of thrombocytopenia, bleeding during airway management and associated risk of haemorrhagic complications. Here we present a case of 25 years young female, 38 weeks of gestation, known case of Evans syndrome and hypothyroidism with bad obstetric history who was successfully managed with spinal anaesthesia and multidisciplinary team approach.

KEYWORDS :

INTRODUCTION

- Evans syndrome is a rare autoimmune condition that presents with two or more cytopenias, which commonly includes
  - o Autoimmune hemolytic anemia (AIHA) and
  - o Immune thrombocytopenia (ITP),
  - o With or without immune neutropenia.
- It can be primary (or idiopathic) or secondary (i.e., associated with an underlying disorder such as systemic lupus erythematosus (SLE), common variable immunodeficiency (CVID), etc.
- The great majority of patients with Evans syndrome have a chronic relapsing course.
- Association of Evans syndrome with pregnancy is very rare. It is diagnosed with a full blood count film and a Coombs test.
- It runs a more benign course in pregnancy than in non-pregnant state (notably neutropenia does not occur) and very often resolves post-delivery.
- Treatment options during pregnancy are further limited due to concerns of teratogenic effect of pharmacological agents.
- The fetal outcome may be less favourable: a minority of fetuses are affected by transplacental passage of antibody and have a significant morbidity and mortality.

CASE REPORT

- 25 year female, G3P1A1IUFD1, 38 weeks of gestation
- Known case of Evans syndrome diagnosed 5 years back
- History of 2 pregnancy losses (G1- spontaneous abortion at 1.5 months in 2017; G2- IUFD at 7 months in 2019 - ?PIH, MRI s/o PRES)
- Known hypothyroidism since 4 years on T. Thyronorm 25 microgram OD.
- She had history of DVT and pulmonary embolism 2 years back for which she was on mechanical ventilation for 7 days and had developed AKI which got resolved after 3 dialysis.
- History of 6 PCV and 10 RDP transfusions in last 5 years.
- Posted for emergency LSCS i/v/o failure of induction of labour.
- She was doing regular hematology follow up for Evans syndrome and was on T. Prednisolone, Inj Rituximab(8 cycles in 2019 and 2020) and Mycophenolate Mofetil.
- At the time of surgery she was on T. Prednisolone(2.5mg OD) and Inj LMWH (0.6 mg BD).
- Inj LMWH was stopped 24 hours prior to surgery.

Investigation And Pre-op Evaluation

Complete Blood Count-

	12/7/21	24/8/21	6/9/21	7/11/21	9/12/21	7/2/22 (present)
Hb (gm%)	11.4	10.3	9.0	9.1	9.4	8.7
WBC	9570	9630	9310	9470	9240	8370
Platelets	2.95	3.50	3.12	2.97	2.58	2.49

- Direct coombs test- Positive
- PT/INR – 13.0 sec /0.96
- FBS – 85 gm%
- PLBS – 112 gm%
- TSH – 1.26 mIU/L
- LFT, RFT, S. ELECTROLYTES – WNL
- ECG – WNL
- GENERAL EXAMINATION : Pt conscious, cooperative, obeying commands.
- Moderately built and nourished.
- Ht- 150 cm, Wt- 55 kg, BMI- 24.4 kg/m2
- No PICCLE. Afebrile.
- VITALS- P-95/min, BP- 125/80 SP02- 99% ON RA.
- Airway examination & systemic examination- WNL

Intraoperative Management

- Starvation confirmed and written informed high risk consent taken.
- Adequate availability of blood and blood products was ensured.
- Patient taken inside the OT after preparation.
- Standard ASA monitoring devices applied.
- 2 wide bore intravenous access were secured and preloading done with 300 ml RL.
- Aspiration prophylaxis was given along with Inj Hydrocortisone 200mg i.v.
- Spinal anaesthesia was administered under all aspectic precautions with 25G Quincke's spinal needle using Inj Bupivacaine heavy (0.5%) 2 cc + Inj Fentanyl 10 microgm at L3-L4 level by midline approach.
- Sensory and motor blockade were achieved till T6 level.
- Procedure lasted for 1.5 hours with stable hemodynamic and urine output 150 ml and blood loss of 400 ml + placenta.
- Post operative period was uneventful and patient was discharged POD-4 and was on follow up with haematology department.

DISCUSSION

- Anaesthetic 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.

- The decision to proceed with neuraxial analgesia or anesthesia in parturients with thrombocytopenia, receiving anticoagulation drugs requires careful consideration, to be individualized and decision making should be based on careful risk-benefit analysis.
- Emphasis should be given on the evaluation of platelet counts before and during pregnancy, bleeding history and possible underlying disorders of hemostasis.
- Strict asepsis must be maintained as these patients are at intrinsic susceptibility and also on immuno-suppressants.
- Perioperative close monitoring of glycemic control and blood loss should be done.

CONCLUSION

- This case was successfully managed under regional anaesthesia despite of autoimmune hemolytic anaemia i.e Evan's syndrome.
- We recommend the importance of multidisciplinary team approach consisting of hematologist, gynaecologist and anaesthesiologist for successful outcome of the patient.

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