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Anaesthetic management of systemic Lupus Erythematosus - Case series

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Abstract

Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune connective tissue disorder. SLE can propose paramount challenges to the anaesthesiologist due to coagulation abnormalities, multiple organ damage and complex treatment regimes. In SLE patients, complications flare up during pregnancy, another important issues includes cardiac involvement, coronary vasculitis, anaemia, thrombocytopenia, thromboembolic phenomena, neurologic involvement and airway manifestations leading to difficult airway. Thus multidisciplinary, individual based approach is utmost important in perioperative management. The present series discusses anaesthesia challenges of Systemic Lupus Erythematosus patients posted for various surgeries.

Keywords: Systemic lupus erythematosus, pregnancy, multisystem involvement, ovarian cyst, cerebral bleed.

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Introduction

SLE is autoimmune disease with multisystem involvement leading to multiple organs and tissues damage by autoantibodies and immune complexes [1]. Magnitude of organ dysfunction is to be noted and an individual approach should be planned on basis of extent of involvement of various systems, ongoing medications and laboratory investigations [2]. People of all genders, ages, ethnic groups are at high risk but majority (90%) patients are women of reproductive age [3]. Incidence and prevalence of SLE varies with community and tribes, it can be predicted as 1/1000 with female to male ratio of 10:1, with peak age of onset 15 to 40 years. Main pathology in patients with SLE includes antinuclear, anti ds-DNA and anti-histone antibodies. American College of Rheumatology criteria are used to draw the diagnosis of SLE. At least 4 out of 11 criteria must be present for diagnosis, which includes photosensitivity, characteristic facial malar rash, discoid rash, serositis (pericarditis and pleuritis), arthritis, renal involvement, neurologic involvement (seizures and psychosis), immunologic phenomena like hemolytic anemia, leucopenia and thrombocytopenia [4]. SLE flares up during pregnancy and will lead to many complications, 33% will have preterm birth 33% may require caesarean section and more than 20% of patients may develop PIH [5]. The risk of preeclampsia is also increased in patients with SLE compared to women without lupus. Risk factors for preeclampsia includes pre-existing hypertension, presence of antiphospholipid antibodies (APL) and nephritis. It is difficult for clinician to differentiate between preeclampsia and nephritic flare as both conditions presents similarly with oedema, hypertension, proteinuria and resulting deterioration in renal function [6]. Here we present anaesthesia challenges of SLE cases during various surgeries.

Case 1

30 years old female patient, G2P1L1 came to the hospital at 38 weeks of gestation, diagnosed with SLE since 10 years and was on treatment for the same. Patient was diagnosed with hypothyroidism at 3rd month of pregnancy and was on treatment for the same. Currently, patient presented with diagnosed preeclampsia from some other practitioner and was admitted for LSCS.

Patient was diagnosed with SLE when she was 19 years of age, at that time patient had lesions all over the body with itching. A complete autoimmune assay was done and SLE diagnosis was confirmed and treatment started with high dose of steroids, but she took the treatment irregularly and treatment was stopped 2 years back. The symptoms had aggravated in the 3rd month of pregnancy, with itching all over the body. Restarted treatment and continued till 6th month of pregnancy. Healed lesions were present all over the back (figure 1) sparing the lower spinal spaces, bilateral upper (figure 2) and lower limbs, ears (figure 3). She had systemic involvement, kidneys manifesting as nephrotic range hypoproteinemia (urine protein – 1513.1), hypertension, cough with expectoration (mucoid white sputum), blackening of nails as a result of a previous episode of Reynauds, anaemia, hypothyroidism, generalised oedema over body including excessive facial puffiness and bilateral pedal oedema, which subsided significantly after treatment with Inj. Albumin. Patient was taking Hydroxychloroquine 200mg HS, Omnacortil 10mg BD, later increased to 60mg BD since last 1 month after nephrologist opinion, patient received Inj. Solumedrol for 3 days,25 days back, Tab. Thyrox 125mcg OD, Tab. Labet 100mg BD, Tab. Nicardia was added 1 week back 5mg BD, Tab.Ecosprin 75mg OD, Inj.LMWH 40 mg SC OD stopped >24hrs before surgery. Following an opinion with the respiratory medicine physician for cough with expectoration, she was started on Inj Deriphyllin 2cc iv TDS, Duolin nebulisation and Budecort nebulisation. Peripheral smear revealed mild anisopoikilocytosis with mild hypochromia few microcytes. Laboratory investigation revealed Complement C3- 28.8mg/dl (75-135), Antinuclear antibodies (ANA) were positive (1:320 end point titre), PFT – FVC 67%,

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FEV167%, MVV 73%. 2D echo was normal. Chest x-ray showed normal fields with prominent bronchovascular markings.

Pre – operatively, patient was advised to continue steroids till day of surgery with antihypertensives, thyroxine and nebulization with Budecort and Duolin before the surgery. Counselling done with risks explained to the patient and relatives. Intra operatively, patient was given Inj. Hydrocortisone 100mg and Inj. Dexa 8 mg before starting induction. Premedication given was Inj. Ondansetron 4mg IV Inj. Ranitidine 50mg IV and Inj.Metoclopramide10mg IV. Pre loading done with 500ml RL. Under all aseptic precautions, SAB was achieved with patient in lateral position by midline approach using a 25G spinal needle at L2-L3 interspace. Inj.Bupivacaine 0.5% heavy, 1.5cc with 25mcg Inj. Fentanyl was given intrathecally after free and clear flow of CSF. Patient made supine immediately after the injection, and the dermatomal level was assessed. T6 dermatomal level achieved. Oxytocin 20 IU was given through the running RL drip to assist placental delivery and uterine contraction. Vitals were monitored, stable throughout the surgery. Urine output was monitored. Post operatively, patient was shifted to recovery unit, block level while shifting was T10. Patient was shifted to obstetric ICU for next 72 hours and then shifted to ward and was discharged on 8th day uneventfully.

Case 2

26year old female with CGA 38 weeks came with C/O headache since 5 days, f/b vomiting, disorientation and irrelevant talk since morning as per history given by relatives. No H/O LOC, breathlessness, chest pain, hemoptysis, hematuria, previous seizures, psychosis, fever or any recent travel history. Patient was diagnosed with SLE in 2019 at 3 months of ANC period in her previous pregnancy. She underwent MTP i/v/o SLE and severe preeclampsia (2019). Since then she was started on treatment consisting of tablet HCQ, Omnacortil, Labetalol and Nicardia which she stopped on her own after 2 months. At present patient she is taking only tablet Labetalol since 2019 with poor compliance. Obstetric History: G1 – 2018 – spontaneous abortion ,G2 – 2019 – MTP at 4th month at LMH i/v/o SLE with severe pre eclampsia On examination patient's general condition was poor ,Afebrile, Pallor +, Anasarca +, Respiratory rate was 24/min with PR: 96/min and BP - 170/110 mmHg she was given Inj. Labetalol 10 mg iv then BP reduced to 120/90 mm Hg followed by GTCS , Inj. MgSO4 (Loading dose) [4 mg iv + 5 mg IM in each buttock] + Inj. Labetalol 10 mg IV given , then GTCS subsided after 20 minutes, DTR (N), Pupils – RTL, plantar flexion +, SpO2 – 96% off O2, B/L conducted sounds + on chest auscultation, No tongue bite, Urine protein 3+, patient then shifted for emergency LSCS. According to preoperative investigation results were Hb – 7.6 gm%, Platelets – 1.39 lacs/cumm, HCT – 21.5, Urea – 30 mg/dl, Creatinine – 0.9 mg/dl, Uric acid – 8.7, Na – 140 mEq/L, K– 3.8 mEq/L, Calcium – 6.6 mg/dl, Mg – 6.6, INR – 0.93, Total protein - 4, Albumin - 1.4, Urine routine, microscopy - Blood +, ANA +, Sm-RNP Ab +, Nucleosome +. Pre op high risk and non NBM consent was taken. Wide bore IV secured. Multipara monitors were attached and patient was catheterized. ECG, NIBP, SpO2, urine output were monitored. Ringer lactate was used as a maintenance fluid. Patient was pre medicated with Inj. Pantoprazole 40mg, Inj. Metaclopramide1mg, Inj. Ondensetron 4 mg. Patient induced with Inj. Propofol 100mg + Inj. Succinylcholine 100mg. Intubated with ET tube number 7 under vision over bougie with RSI technique. Tube placement confirmed and Inj. Atracurium 25mg given. Ryle's tube was inserted under vision. Inj. Hydrocortisone 100mg + Inj. Dexamethasone 8mg given. Inj. Fentanyl 100mcg + Inj. Midazolam 1mg + Inj. Pitocin 10IU given IV after delivery of baby. Patient maintained on Isoflurane + O2 + N2O. Intraoperatively vitals were stable. Urine output was 200ml (high colored). BP ranged from 160/90mmHg – 180/100mmHg. 1point PRC was given. Patient given Inj. Lasix 10mg

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towards the end of the procedure. Extubated uneventfully with Inj. Neostigmine 2.5mg + Inj. Glycopyrrolate 0.4mg. Shifted to Obstetric ICU.

Case 3

A 45year old female, weighing 50 kg, presented with pain in left lower abdomen diagnosed as left ovarian dermoid cyst, posted for exploratory laparotomy. She was a diagnosed case of systemic lupus erythematous and seizure disorder when she had fever, fatigue, joint pain and rashes all over the body with history of seizure episodes 22 years ago she was on regular treatment Tab.Omnacortil 10 mg OD, Tab.Sodium Valproate 200mg OD since then. She had her last seizure episode 12 years ago. She had undergone elective LSCS 24 yrs ago with uneventful pregnancy. She had episode of eruptive drug reaction after taking Tab.Nimesulide for fever with skin lesions all over the body 8 years ago and diagnosed with allergy to multiple drugs including Sulphonamides, Tetracyclines, Ibuprofen, Nimesulide, Metronidazole, Quinines, Dapsone and Trimethoprim. She had history of episode of facial swelling, abdominal distention and vomiting 2 years ago for which she was admitted, detected lupus nephritis and treated accordingly. She was detected with hypothyroidism 1 year ago, taking Tab. Thyroxine 75mcg regularly. She was recently diagnosed with hypertension and started Tab. Amlodipine 5mg OD for the same. On examination, patient had generalised oedema with excessive facial puffiness and periorbital swelling, no malar rash visible, decreased effort tolerance and raised BP of 150/100mmhg. According to investigations TLC were raised to 22,600, TSH was very low (0.02) and 2D echo s/o EF 60% with grade III diastolic dysfunction. Other investigations were within normal limits. On airway examination her mouth opening was adequate with MPC grade II with normal neck movements. On lumbar spine examination, interspinous spaces felt well. Pt was thoroughly counselled a day prior about regional anaesthesia and in case of difficulty or failure, alternative need of general anaesthesia with proper consent. All medications continued till the day of surgery. All standard monitors attached. Patient offered sitting position with support and under all aseptic precautions epidural catheter was inserted in L2-L3 space and subarachnoid block was achieved in L3-L4 space successfully in single attempt. Throughout surgery patient was comfortable and hemodynamically stable. Surgeons found multiple bowel adhesions during surgery and colostomy was done. Patient was shifted to surgical ICU for 3 days post operatively. Block level at the time of shifting was T10.

Case 4

A 42year old female, weighing 55 kg, presented with diffuse pain and distension of abdomen, loss of appetite and vomiting episodes since 2 days. She was the diagnosed case of systemic lupus erythematous sine 18 years, when she noticed joint pain, bilateral cheek rashes and fever during her 1st pregnancy. She was on Tab. Omnacortil 10mg OD since then. Patient was posted for exploratory laparotomy. A thorough preoperative checkup was done. Patient's laboratory were as follows: Hb – 8.2 gm%, Platelets – 1.88 lacs/mm^3, HCT – 23.5, Urea – 35 mg/dl, Creatinine – 1.28 mg/dl, Uric acid – 8.7, Na – 137 mEq/L, K– 3.5 mEq/L, Calcium – 6.9 mg/dl, INR-0.89, Total protein -4.4, Albumin -1.4, Urine routine, microscopy - Blood +, ANA +, Sm-RNP Ab +, Nucleosome +. The plan was general anaesthesia, patient counselled for general anaesthesia with written and informed consent. After applying all standard monitors, she was induced with Inj. Propofol 100mg and Inj. Succinylcholine 100mg, endotracheal intubation was done with ET tube no.7.5 under vision successfully. Intraoperatively patient was maintained on isoflurane and intermittent boluses of Inj. Atracurium. Blood loss and urine output was monitored. On exploration, surgeon found out obstruction at distal ileum, resection and anastomosis was done. Patient had episodes of hypotension intraoperatively for which Inj. Noradrenaline infusion was started and

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maintained @4ml/hr. Intraoperative blood loss was 700ml, she was transfused with 2 bags of PRBC. Patient was then extubated after adequate neuromuscular recovery and shifted with Inj. Noradrenaline infusion@4ml/hr to surgical ICU. Patient's vitals were stable and discharged on postop day 7.

Case 5

A 28yr old female, normal vaginal delivery postop day 3 had drowsiness, headache and confusion and raised BP readings in the range of 160/100mmHg to 180/110mmHg. She was already on Tab. Nicardia 20mcg, Tab. Labet 100mg BD was added to control BP. She suddenly become unresponsive on day 3 of delivery and was intubated and kept on ventilator. She is a diagnosed case of systemic lupus erythematosus since 8 years on regular treatment of Tab.Omnacortil 10mg OD since then. Her current pregnancy was uneventful throughout. CT scan report revealed intraparenchymal cerebral bleed. Her GCS was 6/15, Investigations were within normal limits. Emergency decompressive craniotomy was done under general anaesthesia. Postop patient was kept on ventilator with endotracheal tube in situ for 7 days with alternate VCV and SIMV mode. After that weaning trial was given and patient was extubated on 10^{th} post op day. After extubation she was consious, oriented, responding to verbal commands with stable vitals. All the medications were continued postoperatively. Regular follow up done for 1 month and then discharged home.

Figure 1: Healed lesions present; Figure 2: all over the back; Figure 3: upper limbs Ear

Discussion

SLE is a autoimmune disease, with complex pathogenetic mechanisms which can present at any age, most commonly observed in women in the reproductive age group. It is prone to relapses and remissions. SLE diagnosis is challenging and is mainly based on a clinical presentation. The laboratory and clinical criteria for diagnosis of SLE is determined by American College of Rheumatology which are 95% specific and 85% sensitive [7]. It is important to make sure that the diagnosis of disease is appropriately done before deciding the plan of anaesthesia for the proposed surgery.

Diverse patterns of auto-antibodies and multi-organ involvement is seen in SLE. The disease spectrum ranges from minor organ involvement (e.g. cutaneous lesions) to major organ involvement (e.g., renal, nervous system, etc.) which can be life-threatening. Joints are most commonly affected in SLE patients followed by skin and skin lesions are the second most frequent way of disease presentation [8]. Patients having systemic involvement, may manifest as nephrotic range hypoproteinemia, hypertension, cough with expectoration, blackening of

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nails as a result of associated episode of Raynaud's, anaemia, hypothyroidism, generalised oedema over body including excessive facial puffiness and bilateral pedal oedema.[9]

SLE flare up during pregnancy and pose challenges in distinguishing from disease manifestations to physiologic changes related to pregnancy. Thus, multidisciplinary approach with close obstetric, neonatal and medical monitoring is important to optimize both maternal and fetal outcomes [10]. Different organs may present variable responses to pregnancy; renal and hematologic flare ups are more common than musculoskeletal flares. Clowse ME *et al.* [10] carried a study to determine the risk of rare complications during pregnancy for women with SLE. Maternal mortality was 20-times higher among women with SLE. The risks for thrombocytopenia, infection, thrombosis and transfusion were 3 to 7-fold higher for women with SLE. Pregnant lupus patients had a higher risk for preeclampsia, preterm labor and LSCS than other women. Antiphospholipid antibodies(APLA) are present in many patients with SLE; few patients may develop obstetric or thrombotic complications related to Antiphospholipid syndrome.[11].

Anaesthesia management of SLE patient should focus on multiple organ involvement, deranged coagulation system and airway involvement. Young females with SLE are more prone for coronary artery disease even in the absence of any risk factors. Cardiac involvement can result in myocarditis, pericarditis while valvular lesion characteristically known as Libman-Sacks endocarditis. Coronary vasculitis and accelerated atherosclerosis lead to the high prevalence of ischemic heart disease in these patients. Patients are predisposed to fatal intraoperative events like myocardial infarction. Rhythm and conduction abnormalities are common in these patients. Hence 5 lead ECG and invasive blood pressure monitoring becomes important during major surgeries.

Manipulation of airway needs special care due to possibility of cricoarytenoid arthritis, laryngeal oedema, vocal cord palsy and atlanto-occipital joint subluxation resulting in unanticipated difficult airway. These patients are prone for subglottic stenosis even after a short duration of intubation and in extreme cases they may require tracheostomy. It is safer to avoid intubation using fibreoptic bronchoscope or supraglottic airway devices in these patients.

Anaemia and thrombocytopenia are commonly seen in SLE patients. The presence of lupus anticoagulant is related with prolonged partial thromboplastin time but the risk of bleeding is rare, and thus regional anaesthesia can be practiced safely. The presence of APLA increases risk of venous and arterial thrombosis for which indefinite anticoagulation with warfarin is required to keep INR in between 2 and 4. Bridging therapy is indicated in patients who are high risk for developing thromboembolism. In patients receiving anticoagulants, performing regional anaesthesia should consider benefit and risk as postoperative reintroduction of anticoagulants is necessary. Neurological manifestations in SLE may vary from headache, seizures, stroke to even demyelinating disease. Pulmonary involvement includes acute lupus pneumonitis, diffuse alveolar haemorrhage and interstitial lung disease. Renal involvement is manifested by hypertension, proteinuria and renal insufficiency. General principle for renal protection should be followed even if serum creatinine and urine analysis is normal. Majority of these patients are on prolonged steroid therapy, and it can result in hypothalamic pituitary axis suppression. Patient with cushingoid features on chronic high-dose steroid therapy are more prone for adrenal suppression. Antibiotic prophylaxis should be given as steroids, and immunosuppressant therapy lowers the immunity. Patients receiving immunosuppressant could response differently to anaesthetic drugs. Cyclophosphamide induced plasma cholinesterase inhibition can prolong the action of succinylcholine. Drugs excreting by the renal route should be used judiciously. Normothermia should be maintained as these patients are more prone for Raynaud's phenomenon.

We conclude that anaesthesia management of these patients should be based on a careful

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assessment of multiorgan involvement, hypercoagulable state and perioperative medications [12][13].

Conclusion

The detailed understanding of the pathophysiology of SLE, extent of systemic organ involvement, flaring up of SLE during pregnancy are important consideration for anaesthesia management of these cases.To conclude, judicious perioperative planning and multidisciplinary team approach results in a favourable outcome in patient's with systemic Lupus Erythematosus..

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