



Successful Management Of A Parturient With HELLP Syndrome In The Setting Of Myasthenia Gravis- A Therapeutic Challenge

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Abstract

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Introduction

A Myasthenia gravis is an autoimmune disorder in which the autoantibodies are directed against the post synaptic acetylcholine receptors. In a pregnant patient Myasthenia gravis is found to have a variable effect and myasthenic exacerbations are common in pregnancy especially in the first trimester and towards the puerperium.

Life threatening acute respiratory failure requiring mechanical ventilation may occur due to myasthenic crisis(1). HELLP syndrome is a life-threatening condition which sometimes occurs during pregnancy. The co-occurrence of severe Pre-eclampsia complicated by HELLP syndrome and Myasthenia gravis is extremely rare and mandates strict management strategies to achieve a successful maternal and perinatal outcome. As the literature about this combination is very rare(2), this unique condition poses a large therapeutic challenge to clinicians. The prime issue in decision involves restricted use of anti-hypertensive medications and analgesics

Case Report:

A 35-year-old G2P1L1,29+1 weeks of gestation was referred to obstetric emergency casualty for management of high blood pressure. On arrival patient had HR- 90/min BP-210/110 mmHg and Spo2-98 % at room air. Patient was a known case of Generalized Myasthenia gravis diagnosed six years

ago. She underwent thymectomy in 2017 and was on treatment with Tab Pyridostigmine 60 mg twice a day since then.

She is also known case of hypothyroidism and on treatment with Tab thyroxine 25 mcg once a day.

Patient was shifted immediately to ICU for further management without any delay. Despite initial bolus doses of intravenous labetalol managed by obstetricians, blood pressure was still uncontrolled and anesthesia reference was sent to manage hypertensive crisis. Patient was assessed by our anesthesia team and an invasive arterial line was secured and initially bolus doses of hydralazine 10 mg was administered. Since the blood pressure was high despite bolus doses of hydralazine, Inj.Nitroglycerine infusion was started at rate of 0.3 mcg/kg/min and titrated accordingly.

On day 2, Tab levetiracetam was initiated for seizure prophylaxis. The plan of termination of pregnancy was decided by obstetrician team as the woman developed impending eclampsia. Induction of labor was started. Since the complete hemogram reported thrombocytopenia and baby had severe growth restriction weighing 800 grams, epidural analgesia was deferred. The Criteria for Diagnosis of HELLP syndrome was made. Inj.Nitroglycerin was continued and titrated. Blood pressure was at the range of 150-

160/100-110 mmHg, Urine output was maintained at 50-75 ml/hour.

On day 3, baby was delivered vaginally and was shifted to NICU in view of prematurity and very low birth weight. Inj. Nitroglycerin infusion was tapered and stopped on the same day. Tab. Enalapril was started on postnatal day 1. Patient discharged on postnatal day 4.

Discussion:

Myasthenic crisis are common in pregnancy and it is mainly due to increase in the blood volume and associated changes in the renal clearance of drugs. Myasthenic patients have a variable course during pregnancy among which most of them shows worsening and remaining patients shows no changes with few patients showing improvement(1). Myasthenia Gravis and severe Pre-eclampsia complicated with HELLP syndrome occurs rarely in a co-existing manner. Also, the management and treatment of one disease may worsen the other disease. There are many factors which might influence the myasthenic crisis in a patient during pregnancy and it includes pathophysiological – like stress response, hypo/hyperthyroidism hypo/hyperthermia and pharmacological- like certain antibiotics, Beta blockers, Calcium channel blockers, Magnesium sulphate and anesthetic agents like inhalational agents, neuro-muscular blocking agents(2).

Ideally, patients with Myasthenia gravis should be optimized with their myasthenic symptoms prior to pregnancy. When patient with Myasthenia gravis develops Pre-eclampsia superimposed with HELLP syndrome, as in our patient the main treatment goals is to treat the Pre-eclampsia and control the high blood pressure of the patient. But the problem occurs in treating the high blood pressure. Beta blockers like

Labetalol and other drugs like Nifedipine which were used to control high blood pressure in Pre-eclampsia patients cannot be used in the setting of Myasthenia gravis as chance of exacerbations of Myasthenic crisis is present(2). The same reason holds for initiation of Magnesium sulphate for seizure prophylaxis. Also, Magnesium sulphate impairs already slowed nerve-muscle contractions at the neuromuscular junction through competitive inhibition with calcium. Due to these reasons, drugs like hydralazine can be used to control the blood pressure but, in our patient, even after the bolus dose of hydralazine blood pressure was poorly controlled, So Inj. Nitroglycerin was started as an infusion intravenously to control the blood pressure and we have achieved it. The patient was discharged on postnatal day 6.

Conclusion:

Parturient with HELLP syndrome in the setting of Myasthenia gravis should always be managed by a team of Obstetricians, Anesthesiologists, and Neonatologists in a tertiary care centre(3). Medications need to be considered carefully to avoid problematic interactions and unintentional exacerbation in these patients.

References:

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