Caesarean Section in A Patient with Myasthenia Gravis: A Challenging Anaesthesia

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Abstract

Myasthenia Gravis (MG) is an acquired, autoimmune disorder affecting neuromuscular junction presenting with easy fatigability, progressive weakness, diplopia, difficulty in speaking and swallowing and even ventilator failure in severe cases. During pregnancy the disease may go into remission or may exacerbate at any time during first, second and third trimesters or postpartum period. We are reporting a case of a 32 years old primi gravida, a known case of MG, who underwent emergency caesarian section due to postdated pregnancy. Both the mother and the baby were well managed in the perioperative period.

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Introduction

Myasthenia Gravis (MG) is an acquired, autoimmune disorder affecting neuromuscular junction presenting with easy fatigability, progressive weakness, diplopia, difficulty in speaking and swallowing and even ventilator failure in severe cases. During pregnancy the disease may go into remission or may exacerbate at any time during first, second and third trimesters or postpartum period. Although plenty of literature is available regarding the anaesthetic management of such patients undergoing thymectomy, but clear cut guidelines for those undergoing labor and caesarian delivery are still lacking.

Case report

A 32-year-old primi gravida, a diagnosed case of MG for 1 year. The diagnosis was established after detection of antibodies against acetylcholine (Ach) receptor in patient's serum using radio-immunoassay. A repeated nerve stimulation (RNS) test also supported post synaptic neuromuscular transmission defect. She was under treatment with oral pyridostigmine and prednisolone. After the diagnosis, she did not experience any exacerbation of the disease process and was well controlled on oral pyridostigmine 60

mg four times daily and prednisolone 12.5 mg on odd days and 10 mg on even days.

She reported to our hospital and clinical examination and laboratory parameters including thyroid function tests and ECG were within normal limits. Emergency caesarian section was scheduled due to postdated pregnancy.

A subarachnoid block was instituted with 2.0ml of 0.5% hyperbaric bupivacaine in L3-L4 interspace using 25G Quincke type spinal needle. No perioperative steroid supplementation was given as it may aggravate the weakness. Her hemodynamic and respiratory parameters remained stable throughout the surgery. A 2.8kg healthy male baby was delivered who cried immediately after birth, Appar score being 9,9,9 at 0,1,5-minute. The newborn was shifted to neonatal ICU for observation. After giving a bolus of 5 IU of oxytocin, an infusion was started. At the end of the surgery Diclofenac suppository of 50 mg per rectally and TAP block was given for postoperative pain management. The mother was also shifted to the surgical ICU for close observation and better management. She was closely monitored for respiration, swallowing and speech to detect any evidence of skeletal muscle weakness. The oral pyridostigmine and prednisolone was started after reappearance of bowel sounds, 4 hours after surgery. On the next day evening patient was shifted to the ward with all stable vital parameters and without any evidence of muscle weakness.

Discussion

MG is most common disorder affecting the neuromuscular junction. ⁽¹⁾ It can present at any age but has bimodal peak of incidence. The first peak is seen in females in third decade of life while second peak occurs in sixth decade mainly in females. The etiopathogenesis is characterized by the presence of anti- Ach receptor antibodies in the serum of 85% patients of generalized MG and 50-60% patients of ocular MG. Seronegative patients have anti-muscle specific kinase (MUSK) antibodies in 10-20% cases. There is antibody mediated destruction of Ach receptors present on the post synaptic membrane with loss of folds and widening of a synaptic cleft.²

Abnormality of thymus is seen in 75% cases of MG of which 85% have germinal hyperplasia and 15% have tumor of the gland. Thymectomy is indicated for thymoma and is recommended in all young myasthenics who have a deteriorating response to anticholinesterase drug. ^{3,4} Thymectomy is more beneficial if done prior to pregnancy. Case reports are present when thymectomy was beneficial even when done during pregnancy. ⁵

MG runs a variable and unpredictable course during pregnancy with one third patients improving, one third deteriorating and one third remaining the same during the period of pregnancy.⁶ The worsening of symptoms has been reported to occur in first trimester by some, ^{6,7} in second trimester by a few,8 while others have reported it in third trimester. 9,10 Our patient remained the same during the perioperative period. It is recommended that anticholinesterase therapy be continued orally throughout the period of pregnancy. However, erratic gastric absorption during labor may necessitate a shift to equivalent intramuscular dosages. Steroids, though safe during pregnancy, should be reduced to minimum effective dose. Other modalities like plasmapheresis and azathioprine can be used in case of an acute exacerbation.¹¹ Magnesium sulfate therapy, if needed due to development of PIH, is contraindicated in myasthenics as magnesium may itself produce neuromuscular block by inhibiting release of Ach from the pre junctional membrane. Although the incidence of obstetric complications and surgical delivery are higher in myasthenic patients, it is not an indication for caesarian section and vaginal delivery is preferred. ¹² In our case, vaginal delivery was planned by the obstetrician, as the disease was well controlled, but had to be taken up for surgical delivery due to postdated.

The neonate born to myesthenic mother may experience a transient myasthenic syndrome presenting weak cry, difficulty in sucking and swallowing and respiratory weakness. There are reports of this syndrome of 21%, out of which 67% developed it within few hours of birth. ¹³ Response to oral anticholinesterase is good and complete recovery occurs in less than 2 months. The atypical form may present as arthrogryposis multiplex genitalia. In our case, the baby did not develop any weakness or respiratory distress nor required any anticholinesterase therapy.

It is important for the anaesthesiologist to assess the extent of respiratory or bulbar involvement and determine frequency and severity of myasthenic attacks. The type and dosage of anticholinesterase drug and other medications should be noted and readjusted to obtain optimal symptomatic relief before labor. In severe disease, pulmonary function tests and ECG should be considered. Thyroid function tests may be undertaken as high incidence of autoimmune thyroid disorders are associated with this condition.

Use of labor analgesia is highly desirable to prevent fatigue and exhaustion associated with expulsion efforts during second stage of labor. Opioids are poor choice for this due to risk of excessive depression especially if bulbar or respiratory involvement is present. Central neuroaxial blockade has been used and recommended by many clinicians for vaginal delivery. ^{10,11,14} In our case, labor analgesia was not used as it was decided to be taken up for CS due to postdated pregnancy.

Both regional and GA have been described by various workers for caesarian delivery in myasthenics. Regional anaesthesia is the better