

Caesarean Section in a Patient with Myasthenia Gravis: An Anaesthetic Challenge

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Abstract: Myasthenia Gravis is an acquired, chronic autoimmune disorder which affects neuromuscular junction presenting with easy fatigability, progressive muscular weakness, diplopia, difficulty in speaking and swallowing. Respiratory muscle paralysis may lead to ventilatory failure in severe cases. Myasthenia gravis is characterized by decreased in functioning of acetylcholine receptors at the neuromuscular junctions due to their destruction by circulating antibodies. In 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 the case of a 24 year old primigravida, known case of myasthenia gravis who underwent caesarean section and developed muscular weakness on first postoperative day. Her baby also had a weak cry, hypotonia and tachypnoea. Baby was shifted to neonatal intensive care unit with continuous oxygen supplementation. Baby was kept under close observation and recovered well after one week. Both, the mother and the baby were managed in intensive care unit and responded well.

Keywords: Myasthenia Gravis, fatigability, postoperative, chronic, autoimmune disorder

INTRODUCTION

Myasthenia gravis is an acquired chronic autoimmune disorder caused by decrease in functioning of acetylcholine receptors at the neuromuscular junctions due to their destruction by circulating antibodies. The origin of these antibodies is not known, but a role of thymus gland is suggested. Thymic hyperplasia is seen more than 70% of patients with myasthenia gravis. 10 -15% of myasthenic patients have thymoma [1].

CASE REPORT

A 24-year-old primigravida, a diagnosed case of myasthenia gravis for 3 years, admitted to antenatal clinic of scb medical college hospital for safe confinement. Three years back she had respiratory distress which got relieved on supplementation of oxygen and non invasive ventilation. The diagnosis of myasthenia gravis was made after detection of antibodies directed against acetylcholine (Ach) receptor in patient's serum using radioimmunoassay. She was given oral pyridostigmine and prednisolone. Thymectomy under general anesthesia was done after respiratory distress got cured the perioperative course was uneventful. After thymectomy she was taking oral Pyridostigmine (60mg) thrice daily and prednisolone 10 mg per day till she came to hospital at 30 wks of gestation and was doing well. Clinical examination and laboratory parameters including thyroid function tests and ECG were within normal limits. Emergency cesarian section was planned

for foetal distress at 37 wks of gestation. Caesarean section under spinal anaesthesia was planned. Hydrocortisone 100 mg i.v. was given. Preloading with 1000 mL of lactated Ringer's solution was done and subarachnoid block was given with 2.0 mL of 0.5% hyperbaric bupivacaine and 25 µg fentanyl in L3-L4 interspace using 25G quincke spinal needle. HR, SBP, DBP, MAP, SPO2 and respiratory rate remained stable throughout the surgery. A 2.8 kg healthy male baby was delivered whose, Apgar score being 7, 8, 9 at 0, 1, 5 min. 5U of oxytocin in infusion was started. The newborn was shifted to neonatal ICU for observation. The mother was monitored in intensive care unit for any difficulty in respiration, swallowing and speech. Neostigmine 0.5 mg was given intramuscularly 4 h after the surgery. Oral pyridostigmine and prednisolone started after reappearance of bowel sounds. Postoperative analgesia was achieved with intravenous paracetamol. On first post-operative day, she complained of generalized weakness of whole body particularly of neck and limbs. Gradually ptosis, diplopia, dysarthria developed and patient became dyspneic, and felt difficulty in getting up from bed. The Spo2 dropped to 90% and patient became tachypneic. O2 was administered through mask at 5 l/min. Arterial blood gas analysis showed P_aO₂ of 80 mm Hg and P_aCO₂ of 45 mmHg. Dose of pyridostigmine was increased to 120 mg three times a day. She responded well to the treatment. Her weakness improved gradually over 4 days. In the neonatal ICU, the baby developed tachypnea and hypotonia with poor sucking

and swallowing on second day of her life. ABG showed a P_aO_2 of 75 mmHg and P_aCO_2 of 35mmHg on room air. Oxygen supplementation was sufficient to stabilize the condition with regular suctioning.

DISCUSSION

Myasthenia gravis is an autoimmune, chronic disorder in which muscle weakness is caused by circulating antibodies that block acetylcholine receptors at postsynaptic neuromuscular junction [1]. An estimated 70- 80% of functional acetylcholine receptors are lost, thereby increasing sensitivity to non depolarising muscle relaxants. Women of 20-30 years of age are most often affected. Men are affected commonly after 60 years of age [2]. The hallmark of this disease is muscle weakness and rapid exhaustion of voluntary skeletal muscles with repetitive use. Ocular and pharyngeal muscles are more commonly affected resulting in ptosis, diplopia and dysphagia [2]. In most of the cases weakness of eye muscles are affected first. In myasthenic crises paralysis of respiratory muscles occurs which may require mechanical ventilation. The crises may be triggered by any of the factors like infection, fever, pregnancy or emotional stress [3].

An abnormality of thymus is seen in 70% of cases of myasthenia gravis. Thymectomy is indicated for thymomas & is advised in all young myasthenics who do not respond to anticholinesterase drugs [4]. It is beneficial to do thymectomy prior to pregnancy as was the case with our patient [5]. Complete remission of the disease has been described in approximately 45% of thymectomised patients.

The clinical course of myasthenia gravis and its impact on outcome of pregnancy is unpredictable. Plausche et al noted improvement in 29%, exacerbation in 40% & no change in disease pattern in 31% of myasthenic patients during pregnancy [6]. Worsening of symptoms can occur at any stage during pregnancy but it is more likely during first trimester and first month postpartum. Our patient had disease exacerbation in the early postpartum period resembling the observations of Plausche [6] and Mier *et al.* [7]. It is recommended that anticholinesterase therapy be continued throughout the pregnancy. It improves neuromuscular transmission, suppress the immune system & decrease the circulating antibodies [8]. The dose of corticosteroids should be reduced to the lowest possible level to enhance wound healing and reduce the risk of infection. Plasmapheresis can be safely done throughout pregnancy. It has been recommended as ideal preoperative preparation for myasthenia gravis patient with a vital capacity of < 2L. After plasmapheresis caution should be taken in administration of drugs metabolised by plasma cholinesterase such as succinylcholine, mivacurium, remifentanyl. Immunoglobulins in pregnancy seems to be effective and safe [9]. Vaginal delivery is preferred although emotional stress, physical exertion and fatigue can precipitate crisis. [10]. In our case caesarean section

was planned due to foetal distress. Perioperative management of such patients must include evaluation of extent or severity of myasthenia gravis and optimisation of medical therapy. In severe cases PFT and ECG should be required. Thyroid function test may be undertaken as high incidence of autoimmune thyroid disorders are associated with this condition. Anaesthetic management should stress on managing complications arising because of associated disease, anticholinesterase drugs and plasmapheresis. Myasthenic patients having respiratory and bulbar involvement are more prone to develop respiratory depression with IV opioids. Ester local anaesthetics are not recommended for regional anaesthesia as they are minimally metabolised due to decrease in plasma cholinesterase activity [11, 12]. Amide local anaesthetics which do not require pseudocholinesterase for metabolism are commonly used. It is essential to limit the upper level of block by using lower doses of local anesthetic along with opioids in spinal and graded doses of local anesthetic in epidural anesthesia.

Both regional and general anaesthesia have been administered successfully for caesarean section. General anaesthesia is more appropriate in patients having bulbar and respiratory involvement. Regional anaesthesia is safe in ocular and well controlled generalised disease. Spinal anaesthesia is preferred over epidural as greater quantity of local anaesthetic is needed in epidural block which can precipitate muscle weakness on absorption [11, 12]. In severe disease with bulbar involvement, general anesthesia with endotracheal intubation is recommended. Schnider *et al.* [11] recommended rapid sequence induction with immediate endotracheal intubation in myasthenic parturients. In severely compromised patients, thiopentone alone may be used as sole anesthetic agent. Depolarizing neuromuscular blockers are better avoided due to resistance. Myasthenic patients are sensitive to the action of non-depolarizing agents. One-tenth to one-fifth of the calculated dose is sufficient to cause complete muscle paralysis. This sensitivity is seen in both seropositive and seronegative patients [12], and is more in generalized than ocular myasthenia [13]. Use of inhalational agents should be minimum as they precipitate muscle weakness. Only low doses of short acting opioids are recommended to provide analgesia after delivery of baby. Reversal should be achieved using incremental doses of neostigmine (0.5 mg) intravenously with the response monitored with a nerve stimulator. Overenthusiastic efforts to reverse may precipitate a cholinergic crisis. In the post-operative period, anticholinesterase therapy should be continued intravenously. Once bowel sounds return anticholinesterase should be given orally. These patients can develop complications in the form of myasthenic crisis or cholinergic crisis at any time during pregnancy, labor or post partum period [14]. Post partum exacerbation of myasthenic weakness is usually common [15]. Hence, the patient

should be monitored in intensive care unit for muscle weakness.

The neonate of this myasthenic mother developed a transient myasthenic syndrome presenting as weak cry, difficulty in sucking and swallowing, and respiratory weakness. Papazian [16] reported 21% incidence of this syndrome among the neonates of myasthenic mothers, out of which 67% developed it within first few hours of birth. In our case, the baby developed weakness on second day of life but responded to Oxygen supplementation with regular suctioning.

CONCLUSION

Anesthesiologist plays important role not only in the anesthetic management of a pregnant myasthenic undergoing cesarean section but also in prevention and management of myasthenia crisis which may be precipitated at any time, especially during the immediate postpartum period. Central neuraxial block is preferred in well-controlled myasthenia and general anesthesia in severe cases of myasthenia gravis.

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