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Citation

S Bansal, D Goel, V Gupta, R Jain, V Singhal. *Pregnancy with Myasthenia Gravis with Bad Obstetric History – A case report*. The Internet Journal of Gynecology and Obstetrics. 2008 Volume 11 Number 2.

Abstract

Myasthenia gravis is an acquired immune disorder characterized clinically by weakness and fatigability of skeletal muscle on exertion. It is twice as common in women as in men and frequently affects women in the second and third decades of life, overlapping with the childbearing years. The incidence of myasthenia gravis with pregnancy is 1 in 20,000. Generally, during pregnancy in one third of patients the disease exacerbates whereas in two thirds it remains clinically unchanged. (1) Acetylcholine receptor (Ach R) antibodies are found in around 85% of the patients. (2) In 15% of seronegative myasthenia gravis patients without Ach R antibodies there are serum factors that increase Ach R phosphorylation and reduce Ach R function. Diagnostic test for myasthenia gravis are antibodies to Ach R, muscle specific receptor tyrosine kinase (MuSK Ab), and striated muscle (Str Ab); edrophonium test, EMG and electrophysiological tests. Supporting clinical observations include the presence of thymoma, autoimmune thyroiditis, SLE, Sjogren's syndrome, rheumatoid arthritis and pernicious anemia. 10-15% Neonates born to myasthenia mothers can develop a transient neonatal myasthenia gravis because of transplacental transfer of anti-AchR antibodies (IgG). In rare cases babies of mother with Myasthenia Gravis or even of asymptomatic mothers develop arthrogryposis multiplex congenita caused by antibodies (Fab) that inhibit the ion channel of fetal Ach receptor while not affecting the adult AchR (3). Patients with myasthenia gravis will have a therapeutic response to various immunomodulating therapies including plasmapheresis, corticosteroids, immunoglobulins, immunosuppressants like azathioprine and thymectomy. We report a case of pregnancy with myasthenia gravis with previous neonatal death but with negative acetylcholine receptor antibodies.

CASE REPORT

A 23-year-old P₁A₀, a known case of myasthenia gravis (fig 1) for last 3 years. Though her Acetylcholine receptor antibodies were negative, thymectomy was done in 2002 following which she developed pneumothorax and cholinergic crisis for which plasmapheresis was done and immunosuppressant, anti cholinergic drugs and corticosteroids were given. She recovered well with a long ventilatory support. In her first pregnancy she continued to be on pyridostigmine and azathioprine. Her antenatal period was uneventful, but in last trimester she had decreased fetal movements but was not investigated. Baby delivered by forceps application in a private nursing home where ventilatory support was not available. Baby was floppy at the time of birth and no other anomaly was found on gross examination. Baby died after 6 hours. Due to respiratory insufficiency probably due to neonatal myasthenia. Autopsy

and muscle biopsy of baby was not done.

She planned her second pregnancy after proper pre-pregnancy counseling by consultation of neurologist and obstetrician and was on neostigmine & corticosteroids throughout her antenatal period. All investigations were within normal range. (Table No.1) Level II scan and fetal echocardiography were normal. Serial ultrasonography showed normal fetal movements, Fetal growth and amniotic fluid. Triple test at 16 weeks & fetal echocardiography at 24 weeks was also normal. Patient was kept on low dose aspirin till 34 weeks of pregnancy as a prophylaxis for PIH.

She went into spontaneous labour at 37 weeks. At the time of admission her vitals were within normal limits. There was no sign of muscle fatigue. On per vaginam examination, cervical os was 4 cm, dilated fully effaced with vertex at -1 station, bag of membranes forming, and pelvis adequate.

Artificial rupture of membrane was done, liquor was clear. Labour was augmented with intravenous syntocinon drip. Antibiotic cover was given and kept on strict bed rest in lateral position with adequate analgesia with tramadol. Labour was monitored carefully and hydrocortisone 100 mg intravenously was given. Pyridostigmine was continued in same dose. Neostigmine 1 ml (0.5 mg) was given IV slowly given in second stage of labour when patient started bearing down. She delivered a healthy male child of 2.78 Kg. Episiotomy was given. Baby cried immediately after birth. Apgar score was 9/10. Total duration of labour after admission to hospital was two and a half hour. Post partum period was uneventful. No ventilatory support was required for mother or baby.

After 3 to 4 hrs of birth baby had slight drooping of milk. She was given intravenous neostigmine in the dose of 0.01 to 0.04 ml /kg. Baby did not require ventilatory support and was given to mother for breast-feeding after 6 hrs. Baby was discharged after 3 days after careful monitoring. At postnatal check-up after 6 weeks, both mother and baby were healthy.

Figure 1

Figure 1: Patient’s photograph showing old tracheostomy scar



Figure 2

Table – 1 Investigations Results

S.No.	Investigations	Result
Routine:		
1	Blood group	B +ve
2	Hemoglobin	11.8%
3	TLC	11200 c/mm ³
4	DLC	N ₇₄ L ₂₄ E ₀ M ₀
5	Urine R M	WNL
6	Blood Sugar: FBS PPBS	78 mg/dl 83 mg/dl
7	VDRL	NR
8	HBS Ag	NR
9	HIV	Non reactive
10	Specific:	
11	Acetyl choline Receptor antibodies	Negative
12	Repetitive Nerve stimulation test	Positive
13	Electromyography	Neurogenic pattern
14	Thyroid profile	Normal range
15	Fetal Echocardiography	Normal scan
16	Alpha Feto protein	67.17 mg/ml - Normal
17	Triple test	Screening Negative

DISCUSSION

The course of disease is unpredictable during pregnancy, however worsening of symptoms occurs more likely during first trimester and postpartum. Complications at delivery are higher (40.9% Vs 32.9%) i.e. fetal distress, preterm rupture of membranes (5.5 % Vs 1.7 %), interventions during delivery (33.9% Vs 20.0%) and Caesarean section doubled (17.3% Vs 8.6%) (4). MG can be well managed during pregnancy with safe and effective therapies.

Anticholinesterase drugs are the mainstay of it, along with corticosteroids and azathioprine. Plasmapheresis and intravenous immunoglobins if required (4). In our patient, cholinergic crisis was managed by plasmapheresis and immunomodulating drugs and pregnancy was successfully managed with neostigmine & corticosteroids with good maternal and perinatal outcome.

During antenatal period patient should be managed in a tertiary care center with regular follow up with neurologist and obstetricians. Physical activity of patient should be kept minimum. Fetal movements, fetal growth and amniotic fluid should be monitored with frequent ultrasound.

During labour patient should be kept on strict bed rest in lateral position. Adequate hydration, antibiotic coverage & adequate analgesia should be given. Caesarean section is recommended only for obstetrics reasons. Epidural analgesia is the choice of analgesic in labour. Premature bearing down and prolonged Labour should be avoided. Some medications are contraindicated in labour. These are magnesium sulphate, aminoglycosides, halothane, Propranolol, Cholistin, Polymyxin B, Quinin, Tetracycline, Barbiturates, Trichlorethylene. Ventilator and anesthetist should be

available to manage respiratory insufficiency. Liberal episiotomy and Prophylactic outlet forcep should be applied.

Baby should be monitored for any signs of respiratory insufficiency for upto three weeks. Maternal myasthenia have been associated with the presence of neonatal myasthenia and sometimes-fatal congenital anomalies. Risk of neonatal myasthenia gravis is not related with severity ,duration of maternal disease or antibody titre but can be halved if mother is thymectomized . In the case described above, though mother was AchR negative her both babies were effected.The transmission of the disorder from mother to child even in seronegative myasthenia gravis clearly indicates that there is a circulating serum factor,other than Ach R, probably IgG in SNMG. Recently, IgG antibodies against muscle-specific kinase (MuSK) have been demonstrated in up to 2/3rd of cases with SNMG (5). Moreover, plasma from patients with SNMG also seems to contain a factor, possibly IgM that indirectly inhibits AchR function by phosphorylation of AchR(6)

CONCLUSION

Myasthenia gravis can be successfully managed during pregnancy with a combination of medical drugs with good maternal and fetal outcome. Women with myasthenia gravis

should not be discouraged from conceiving; however they should discuss their plan for pregnancy with their neurological and gynaecologist and should have intensive check up throughout their antenatal periods.

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