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Third caesarean section in patient with myasthenia gravis

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ABSTRACT

Myasthenia gravis (MG) is an autoimmune neuromuscular disease, characterised by muscle weakness and fatigability of the voluntary muscles, it affects young women in the second and third decade of life. We report a case of 30 years old multigravida woman with myasthenia gravis submitted to third iterative caesarean section with no adverse neonatal and maternal outcome. The course myathenia is highly variable and unpredictable during gestation and can change in subsequent pregnancies. Delivery specially, through caesarean section is very stressful and may cause severe myasthenic crisis. Although pregnancy and delivery represent particular events, that require more attention in these patients, they are not associated, in most cases, with higher risks of complications compared to normal pregnancy, delivery and postpartum period. According our experiences, in the management of myasthenic pregnant woman is necessary cooperation in a multidisciplinary team between obstetricians, neurologist, anaesthetist, and neonatologist for ensure an optimum outcome.

1. Introduction

Myasthenia gravis (MG) is an autoimmune disorder caused by autoantibodies against the nicotinic acetylcholine receptor on the postsynaptic membrane at the neuromuscular junction and characterised by weakness and fatigability of the voluntary muscles[1]. It has a bimodal peak of incidence with first peak in the second and third decade, overlapping with the childbearing years[2] (incidence in pregnancy 1:20 000)[3] and the second peak in sixth decade.

Myasthenia gravis can be classified according to the age of onset, presence or absence of anti-AChR antibodies, severity according to Osserman's classification, and aetiology (congenital myasthenic syndromes, acquired autoimmune, transient neonatal and drug induced).

The effect of pregnancy on myasthenia gravis is variable. Approximately one third of myasthenic pregnant get better and one third get worse at some time during their pregnancy, while one third do not change. There is usually worsening of disease in the first trimester and an improvement is noted in the third trimester. Complete remission can occur in some patients[4]. Anticholinesterase medicines and corticosteroids are the mainstays of medical therapy of maternal myasthenia gravis and require frequent adjustment during pregnancy due to changing requirements and physiologic changes in absorption and excretion. Thymectomy, plasmapheresis, immunosuppressant drugs, gammaglobulin and ACTH are adjuvants of varying usefulness. The treatment anticholinesterase medications and steroids have not been found to be associated with significant risk for congenital defects. Plasmapheresis has been carried out safely during pregnancy. Myasthenia gravis affects striated muscles and the uterus is a smooth muscle. Therefore, the obstetric complications are not very common during delivery. Only during the second stage of labour when voluntary ''striated'' abdominal muscles are used does myasthenic weakness become noticeable. In studies looking at the effects of myasthenia gravis on pregnancy and delivery, complications of artificial rupture of amniotic membranes with chorioamnionitis, preclampsy and the rate of interventions were noted to be greater in patients with myasthenia gravis.

Regarding the newborns, the incidence of transient neonatal myasthenia gravis has been reported between 9% and 30%[2]. Typical clinical findings of transient neonatal myasthenia gravis are poor sucking and generalized hypotonia. Other manifestations are swallowing and sucking difficulties and mild respiratory distress[5].

We report a case of myasthenia gravis in a woman who was managed successfully through caesarean deliveries in all her pregnancy with good outcome.

Written consent for publication of the present report was obtained from the patients.

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2. Case report

A 30 years old gravida 3 para 2 woman was referred to our hospital at 38 weeks of gestation, with an obstetric history of previous two caesarean sections. She was diagnosed to have myasthenia gravis type II A according to Osserman's classification used by Myasthenia Gravis Foundation of America, since the age of 16 years, with the development of classical symptoms of this disease, she was seropositive for anti-AChR antibodies too. For treatment thymectomy was carried out. Drug therapy consisting of an anticholinesterase agent was initiated. Her condition responded to anticholinesterase treatment and she needed increasing daily dosages over the years. During pregnancies, her pyridostigmine dosage was 90 mg orally every 6 hours which was maintained throughout the three trimesters of gestation, no alteration in dosage of medicine was necessary during and after all pregnancies.

Her first pregnancy at age 22, ended at 39 weeks of gestation, she delivered a female baby, weighing 2.630 kg, through caesarean section, under spinal anaesthesia, performed due to breech presentation. The neonate suffered from transient neonatal myasthenia gravis, this baby presented sucking difficulties and mild respiratory distress.

In the second pregnancy, at age 25, ended 38 weeks of gestation, did not change the clinical status compared with before pregnancy, a caesarean delivery was performed under spinal anesthesia. The weigh of the male newborn, was 2.930 kg, he remained asymptomatic.

In this pregnancy, careful monitoring was performed closely by obstetric and neurologist, after an anaesthesiology assessment, elective caesarean section was carried out on term, under spinal anaesthesia, the cholinesterase level was 4 370 U/L. During delivery was required the preventive presence of Neonatal Intensive Care Unit, for a possible respiratory distress. A male baby weighing 2.860 kg was born, with an APGAR scores of nine at one minute, and ten at five minutes, without complications. No congenital anomalies were detected in any of the newborns. The patient continued her normal oral therapy before and after the operation (Pyridostigmine, 360 mg daily). During pregnancies her conditions remained stable. She had an uncomplicated postoperative and perpueral course.

3. Discussion

Myasthenia gravis is an acquired relapsing and remitting autoimmune disorder associated with acetylcholine receptor (AChR) deficiency at the motor end plates, caused by complement fixing antibodies, it has an incidence rages from 1:10 000 to 1:50 000[2].

Thymectomy is a standard procedure for enlarged thymus gland. Removing a major source of antibody production clinically improves myasthenia gravis.

Literature describing the clinical course of pregnant myasthenic woman mostly consists of single case report and case series[4].

In pregnancy, the course of myasthenia gravis has been reported to be highly variable and unpredictable in the antepartum period. Outcome in a previous pregnancy is not predictive of what the future course might take in a subsequent pregnancy[6].

In myasthenic pregnants receiving anticholinesterase therapy there is a higher possibility of spontaneous abortion or premature labour, because anticholinesterase drugs represents a potential risk for stimulation of the uterine cholinergic receptors leading to an increase in uterine tone and contractions. Myasthenia gravis has not been shown to affect the course of labour and is not, in itself, an indication for caesarean section. However, it would be undesirable for labour to be prolonged and exhausting. Other author conclude that there were no statistically significant differences in the risk of having preterm, LBW, SGA infants and caesarean deliveries between women with and without myasthenia ravis[7].

Regarding newborns the incidence of transient neonatal myasthenia gravis, has been reported between 9% and 30%. The occurrence of neonatal myasthenia gravis does not correlate with either maternal disease severity or anti-AChR antibody titer and the severity of MG in the in the infants is variable and does not correlate with neither the severity of maternal MG during pregnancy of if it is the first or subsequent pregnancy. The clinical manifestations more common, are generalized hypotonia, sucking difficulties and mild respiratory distress.

Respiratory distress requiring assisted mechanical ventilation can occur in severe cases. Complete recovery is expected in less than 2 months in 90% of patients and by 4 months of age in the remaining 10%[2].

The literature on myasthenia gravis and pregnancy is rather limited. Although pregnancy, delivery and breastfeeding represent particular circumstances in these patients, they are not associated, in most cases, at higher risks of complications compared to normal pregnancy, delivery and postpartum period[8]. According our experiences, in the management of myasthenic pregnant woman is necessary a cooperation in a multidisciplinary team between obstetrician, neurologist, anaesthetist, and neonatologist for ensure an optimum outcome.

Conflict of interest statement

We declare that we have no conflict of interest.

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