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Myasthenia gravis: a case report

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Abstract:
Myasthenia gravis in pregnant women requires multidisciplinary management (neurologist, obstetrician, anesthetist and pediatricians) due to the risk of maternal myasthenia gravis and the risk of neonatal myasthenia gravis. We present the case of an 18-year-old primigravida with myasthenia gravis for which an immunosupressant and pirydostigmine were administered even during the preconceptional period and first trimester. The patient exhibited several relapses during her pregnancy none of which had an impact on her respiratory system. The labourand delivery were closely monitored and came to pass with ease. The post partum period was uneventful for the mother and newborn.
Regular neurological monitoring during pregnancy and the postpartum period should be implemented with the aim of early detection of relapses and the establishment of specific treatments for an optimal management of any situation

Key Word*:Myasthenia gravis, pregnancy, neonatal myasthenia*

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I. Introduction

Myasthenia gravis is a chronic autoimmune neurological disease that develops in flare-ups/remissions. It mostly affects young women, of childbearing age, therefore, the association of pregnancy and myasthenia is not exceptional. The prevalence is 0.25–2 affected individuals per 100,000. There are two peaks in the prevalence of the disease, between 20 and 30 years of age with predominance of women (SR 2/1) [1-3], and after 40 years when the sex ratio becomes 1/1 [2].

It is therefore necessary to know the distinctive features of these pregnancies to ensure an optimal monitoring, in complete safety for both mother and child. Pregnant myasthenic patients are exposed to more frequent relapses [3].As for the new born, the major risk is neonatal myasthenia gravis which may require the monitoring of the child in intensive care [3].

II. Case report

We present the case of an 18-year-old, primigravida , diagnosed with myasthenia gravis at the age of 14, etiologic exploration revealed the presence of a thymic hyperplasia for which the patient underwent a thoracoscopic thymectomy. immunosuppressants and an antagonist to cholinesterase were also introduced in order to control her symptoms and improve her quality of life.

Due to the below average standard of living and the unplanned nature of the pregnancy, the patient could not access a pre conceptional consultation, therefore no therapeutic adjustments were undertaken, and folic acid/ iron supplementation was not administered.

The patient benefited from 4 prenatal consultations starting at 16 weeks, a standard prenatal screening came back negative, and 3 ultrasound examinations that showed no morphological or growth abnormalities.

In addition, the patient benefited from a regular follow up by her neurologist, who determined the necessity of maintaining Azathioprine 50mg x3/day and pyridostigmine 60mg x4/day treatment. Attempts at decreasing the dosage of the immunosuppressant resulted in mild relapses consisting mainly of fatigability, ptosis, diplopia, dysphagia, dysarthria and fatigable chewing of solid foods. Such relapses were also seemingly triggered by stressful situations and/ or infections.

At 39 weeks, the patient consulted the emergency room for uterine contractions, the clinical assessment found an asymptomatic patient in terms of muscle strength (none of the symptoms cited earlier were present) normo-colored conjunctiva, normotensive, afebrile and the obstetrical assessment uncovered a patient in the latent phase of labor.

At 3cm, epidural analgesia was administered. The partograph was closely monitored showing a smooth progress of labor, and the vaginal delivery required no instrumentation, giving birth to a healthy male newborn Apgar 10/10, weighing 3200g at birth, the initial pediatric assessment was normal, nevertheless, the

new born was transferred to the neonatal care ward for monitoring given the risk of neonatal myasthenia gravis of which no signs were detected.

The postpartum period proved uneventful with the patient remaining asymptomatic under treatment.

III. Discussion

Myasthenia gravis results from the blockage of transmission at neuromuscular synapses by anti-nicotinic acetylcholine receptor antibodies. Smooth muscles (including the uterus) that have muscarinic acetylcholine receptors are not affected by anti-nicotinic acetylcholine receptor antibodies the duration of labor remains therefore unaffected. Some authors have recommended instrumental extraction to reduce expulsive efforts and reduce maternal fatigue (4). Concerning our patient, there was no need for instrumental extraction as the labor and delivery proceeded smoothly and without delay.

Myasthenia gravis is a chronic disease which progresses in relapses and remissions. It usually results in increased fatigue on exertion, swallowing disorders, visual disturbances with ptosis, or breathing difficulties that may require ICU hospitalization in (1).

According to the current literature, the evolution of myasthenia gravis during pregnancy is unpredictable, authors report 1/3 of cases would show no improvement, 1/3 would remain asymptomatic, and 1/3 would present an aggravation with a risk of myasthenic crisis which could be severe, sometimes affecting the respiratory muscles, this complication could happen during the any time of the pregnancy and even during the postpartum(4-5-6). The flare-ups are however favored by certain drugs of frequent use during pregnancy, namely: calcium channel blockers and intravenous magnesium.

Anticholinesterase drugs have no teratogenic effects and should not be discontinued, especially during labor to prevent the risk of decompensation (7).

Epidural analgesia is not contraindicated since pain, anxiety and fatigue can cause decompensation during labor. These three factors are well controlled by the establishment of effective analgesia. Epidural analgesia is therefore clearly a medical indication in myasthenic parturient, because it reduces fatigue, regulates maternal ventilation and above all facilitates instrumental extraction (8).

In terms of the fetal complications of myasthenia, some cases of hypotrophy (20-35%), arthrogripposis by fetal immobility and pulmonary hypoplasia by abolition of respiratory movements have been reported. Hydramnios by failure to swallow even allowed the diagnosis of maternal myasthenia gravis in the case published by Verspick et al. (9). Taking pyridostigmine in high doses would be associated with a risk of fetal microcephaly for patients. However, close ultrasound monitoring with evaluation of fetal vitality is warranted.

Neonatal myasthenia gravis is found in 10 to 20% of cases in the literature, with the symptoms developing in the first 24 hours of life with sucking and swallowing disorders, weak cries or respiratory distress. It is due to the transplacental passage of anti-acetylcholine receptor antibodies, the presence of which does not exceed three weeks in the child's system. The delayed onset of neonatal disease is due to both the elimination of anti-acetylcholinesterase drugs and the drop in alpha-fetoprotein (AFP) levels. AFP has been shown to inhibit binding of the anti-acetylcholine receptor antibody to its ligand [10]. 80% of newborns of myasthenic mothers have circulating antibodies although they are free from neurological signs [11]. Treatment with neostigmine is usually sufficient, combined with intensive care in severe cases.

Antibodies to acetylcholine receptors and anticholinesterase drugs are known to pass into breast milk. These can have muscarinic-like side effects in children [3, 11, 12], without having teratogenic effects. Breastfeeding is controversial, some authors believe that a patient in remission with low levels of antibodies and without dangerous drugs can breastfeed without risk, and that artificial breastfeeding should be preferred in other cases [2-3].

IV. Conclusion

We can conclude that pregnancy can be allowed without restriction if preconceptionally balance is achieved. On the obstetrical level, myasthenia gravis has little impact: labor is a physiological phenomenon. However, the disease can have fetal and neonatal repercussions. Regular neurological monitoring during pregnancy and the postpartum period should be set up with the aim of early detection of relapses and the establishment of specific treatments to stop them. This is why perfect cooperation from a multidisciplinary team (neurologist, obstetrician, anesthesiologist and pediatricians) is necessary.

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