CASE REPORT

Maternal Myasthenia Gravis: A Case Report

Mary Devasiya[1], Akshaya Murali[2], Anitha Santosh[3], Madhu Swetha Sharma[4]

Received on: 08 July 2024; *Accepted on:* 08 August 2024; *Published on:* 23 October 2024

Abstract

Introduction: Myasthenia gravis (MG) is an acquired neuromuscular disorder that presents with clinical weakness and fatigue of the skeletal muscles. Pregnancy can have significant effects on the course of myasthenia gravis. Patients with well-controlled MG prior to conception usually have a benign disease course during pregnancy, although a small number may deteriorate during the postpartum period.

Case description: A 22-year-old primigravida was referred to our hospital for further antenatal management, as she was a known case of MG. Myasthenia gravis was diagnosed at the age of 17, when she developed drooping of eyelids, and weakness of both upper and lower limbs. Acetylcholine receptor antibodies were positive. Her symptoms improved with oral prednisolone and pyridostigmine, but caused bilateral avascular necrosis of the hip requiring core decompression surgery. She further required thymectomy a year later. The patient's myasthenic symptoms were well-controlled prior to conception and she remained asymptomatic throughout pregnancy while continuing pyridostigmine. She underwent an elective cesarean section at 39 weeks of pregnancy due to her prior history of avascular necrosis of bilateral hip and her postpartum period was uneventful.

Conclusion: Pregnancies associated with MG can have a stable disease course if adequately stabilized with drugs or thymectomy prior to pregnancy. Antenatal management by a multidisciplinary approach, along with appropriate immunosuppressants can improve both maternal and neonatal outcomes.

Keywords: Case report, Fetomaternal outcomes, Myasthenia gravis, Neurological disorder.

Journal of South Asian Federation of Obstetrics and Gynaecology (2024): 10.5005/jp-journals-10006-2512

Introduction

Myasthenia gravis (MG) is an acquired neuromuscular disorder that presents with clinical weakness and fatigue of the skeletal muscles.[1] Myasthenia gravis has a prevalence of 126–400 per million in the general population.[2] Myasthenia gravis is an autoimmune disease of the neuromuscular junction (NMJ), which is caused by autoantibodies directed against acetylcholine receptors (Anti-AchR), muscle specific kinase (MuSK) or LRP4 on the postsynaptic membrane.[3,4] These antibodies injure the NMJ, resulting in skeletal muscle weakness. Myasthenia has two clinical forms: Generalized or ocular type. Patients with anti-AChR positive MG often tend to have thymic abnormalities, such as thymic hyperplasia in 60–70% of the cases, and thymoma in 10% of the cases.

Pregnancy can alter the course of MG, but those with well-controlled MG prior to conception usually have a benign antenatal period, although rarely, there may be deterioration in the postpartum period.[3] Flares are more common during the first trimester and the immediately post-delivery period in women with pre-existing active disease. Some people may be diagnosed with MG after experiencing their first symptoms during pregnancy. The condition frequently affects young women between 20 and 40 years of age, and has a high chance of coinciding with pregnancy.[5] The disease course is often unpredictable, with a significant worsening in one-third of the affected women, while the rest stay the same or improve.

Transient neonatal MG (TNMG) is a transitory form of myasthenia which affects 12–20% of newborns delivered to myasthenic mothers.[6] Transient neonatal MG is caused by placental transfer of nicotinic AchR antibodies from the mother. It presents with poor sucking, muscle weakness, weak cry, and drooping of eyelids in a majority of the cases. Rarely, severe respiratory depression can occur. These symptoms resolve by 1–3 months of age, following the breakdown of maternal-derived antibodies.

[1–4]Department of Obstetrics and Gynaecology, St. Johns Medical College Hospital, Bengaluru, Karnataka, India

Corresponding Author: Akshaya Murali, Department of Obstetrics and Gynaecology, St. Johns Medical College Hospital, Bengaluru, Karnataka, India, Phone: +91 9632203900, e-mail: dr.akshayamurali@gmail.com

How to cite this article: Devasiya M, Murali A, Santosh A, *et al*. Maternal Myasthenia Gravis: A Case Report. J South Asian Feder Obst Gynae 2024;16(5):576–577.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

Case Description

A 22-year-old primigravida was referred to the out-patient department of our tertiary care hospital in the third trimester of her pregnancy. She was a known case of MG since 5 years, and was on regular medication throughout pregnancy without features of exacerbations.

Myasthenia gravis was diagnosed at the age of 17, when she developed drooping of eyelids, and weakness of both upper and lower limbs. On detailed evaluation, the neostigmine test was positive, RNS (3Hz), showing decremental response in the function of trapezius and abductor pollicis brevis muscles. Acetylcholine receptor antibodies (Anti-AchRs) were positive. Her symptoms improved with oral prednisolone and pyridostigmine. Prolonged steroid use resulted in bilateral avascular necrosis of the hip (Mitchell Class C) 2 years later, requiring core decompression

surgery. She further required four cycles of lymphoplasmapheresis as well as thymectomy for thymic hyperplasia a year later, following which she went into remission.

The patient's myasthenic symptoms were well-controlled prior to conception and she remained asymptomatic throughout pregnancy while continuing pyridostigmine. Both maternal and fetal monitoring showed no deterioration through any of the routine antenatal visits. Neurological monitoring was carried out on OPD basis and indicated a stable course of disease.

Due to her prior history of avascular necrosis of bilateral hip, the patient could not abduct or flex at her hip joint. She underwent an elective cesarean section at 39 weeks of pregnancy to deliver a live baby girl of 2800 gm. Postpartum period was uneventful. The new-born was screened for TNMG, monitored for muscle weakness and respiratory distress. Both mother and baby were discharged on post-cesarean day 4 in satisfactory condition.

Discussion

Pregnancies complicated by MG may vary in presentation from one woman to another, depending on the prepregnancy stability of the disease. There can be an improvement of symptoms in the second and third trimesters, which has been associated with normative immunosuppressive alterations in late pregnancy. Symptomatic flares are most likely to happen during the first trimester or right after birth.[5]

Treatment options for MG differ based on the underlying causative antibodies and patient symptomatology. Acetylcholinesterase inhibitors (pyridostigmine, neostigmine) are the preferred first-line treatment options and are more effective in patients with anti-AChR antibody, compared with those with anti-MuSK antibody. Patients with MuSK antibodies respond well to high-dose corticosteroids, which are required long-term.[6–8] Immunosuppressants like mycophenolate mofetil are teratogenic and must be substituted with safer drug options 6–8 weeks before planning conception.

Well-controlled myasthenia has no significant negative consequences on pregnancy. Women with MG generally are not at a higher risk of spontaneous abortions or early births. Pregnancies do not affect the long-term course of the disease. The choice of treatment should be tailored to the patient's requirements. Pyridostigmine, low dose steroids, and azathioprine are safe in pregnancy and can be continued. Mycophenolate mofetil, methotrexate, cyclophosphamide, and Rituximab are contraindicated and alternate drug options should be discussed prior to planning pregnancy. Monoclonal antibodies like Eculizumab are rarely indicated when benefits outweigh the risks.[9] Thymectomy should be explored before pregnancy, if generalized MG is positive for anti-AChR antibodies. Pregnancy should be avoided for the first 2 years after the initial diagnosis of myasthenia due to an increased risk of disease deterioration in the early stage.[3,5,10]

Inoue reported a case of MG (MuSK positive) which exacerbated after pregnancy, requiring oral steroid therapy and double filtration plasmapheresis. The baby developed TNMG which spontaneously resolved in a month. It is important to note that women with MuSK-MG can deteriorate during or after pregnancy, and pose a high risk for TNMG.[4,6] Aitken described a case of MG with initial presentation in the third trimester of pregnancy with diplopia and muscle weakness, and identified to be MuSK positive. The patient was managed with prednisolone and plasmapheresis successfully and the baby showed no features of TNMG.[7] Assudani described another case of MG first diagnosed antenatally, managed with azathioprine and prednisolone, with no flare up during or after pregnancy.[11] In the case report described by Berlit et al., maternal MG with myasthenic crisis progressed to respiratory insufficiency in the postpartum period requiring intubation. A multi-disciplinary intensive care approach was used to aid patient recovery.[10]

The severity and duration of MG, as well as adequate treatment prior to pregnancy can have an impact on the antenatal course.[12] Thymectomy before delivery has been associated with improved clinical outcomes.[13] Women with MG require prepregnancy stabilization and routine monitoring of the mother and the fetus by a multidisciplinary team approach including obstetricians, neurologists, neonatologists and rarely intensivist, to enhance pregnancy outcomes and reduce morbidity.

Conclusion

Pregnancies associated with MG can have a benign course if adequately stabilized with drugs or thymectomy prior to pregnancy. Antenatal management by a multidisciplinary approach, along with appropriate immunosuppressants can improve both maternal and neonatal outcomes.

Orcid

Akshaya Murali https://orcid.org/0000-0001-5836-7241

References

1. Téllez-Zenteno JF, Hernández-Ronquillo L, Salinas V, et al. Myasthenia gravis and pregnancy: Clinical implications and neonatal outcome. BMC Musculoskelet Disord 2004;5:42. DOI: 10.1186/1471-2474-5-42.
2. Norwood F, Dhanjal M, Hill M, et al. Myasthenia in pregnancy: Best practice guidelines from a UK multispecialty working group. J Neurol Neurosur Psychiatry 2014;85(5):538–543.
3. Hamel J, Ciafaloni E. An Update: Myasthenia gravis and pregnancy. Neurol Clin 2018;36(2):355–365. DOI: 10.1016/j.ncl.2018.01.005.
4. Evoli A, Tonali PA, Padua L, et al. Clinical correlates with anti-MuSK antibodies in generalized seronegative myasthenia gravis. Brain 2003;126(Pt 10):2304–2311. DOI: 10.1093/brain/awg223.
5. Chaudhry SA, Vignarajah B, Koren G. Myasthenia gravis during pregnancy. Can Fam Physician 2012;58(12):1346–1349. PMID: 23242891.
6. Inoue KI, Tsugawa J, Fukae J, et al. Myasthenia gravis with anti-muscle-specific tyrosine kinase antibody during pregnancy and risk of neonatal myasthenia gravis: A case report and review of the literature. Case Rep Neurol 2020;12(1):114–120. DOI: 10.1159/000506189.
7. Aitken A, Kanitkar S. New diagnosis of myasthenia gravis in the third trimester – A case report. Case Rep Womens Health 2023;38:e00505. DOI: 10.1016/j.crwh.2023.e00505.
8. Meriggioli MN, Sanders DB. Autoimmune myasthenia gravis: Emerging clinical and biological heterogeneity. Lancet Neurol 2009;8(5):475–490. DOI: 10.1016/S1474-4422(09)70063-8.
9. Gilhus NE. Treatment considerations in myasthenia gravis for the pregnant patient. Expert Rev Neurothe 2023;23(2):169–177. DOI: 10.1080/14737175.2023.2178302.
10. Berlit S, Tuschy B, Spaich S, et al. Myasthenia gravis in pregnancy: A case report. Case Rep Obstet Gynecol 2012;2012:736024. DOI: 10.1155/2012/736024.
11. Assudani L, Notwani A. Myasthenia gravis in pregnancy: A case report. Int J Reprod Contracept Obstet Gynecol 2023;12(1):264–267. DOI: 10.18203/2320-1770.ijrcog20223508.
12. Ducci RD, Lorenzoni PJ, Kay CS, et al. Clinical follow-up of pregnancy in myasthenia gravis patients. Neuromuscul Disord 2017;27(4):352–357. DOI: 10.1016/j.nmd.2017.01.021.
13. Su M, Liu X, Wang L, et al. Risk factors for pregnancy-related clinical outcome in myasthenia gravis: A systemic review and meta-analysis. Orphanet J Rare Dis 2022;17(1):52. DOI: 10.1186/s13023-022-02205-z.