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Myasthenia Gravis and Pregnancy: A Case Report

A. Jamaleddine[1], M. Sali[1*], M. Trabelsi[1], H. Asmouki[1], A. Aboulfallah[1], B. Fakhir[1], A. Bassir[1], K. Harou[1], L. Boukhani[1], A. Soummani[1]

[1]Obstetrics and Gynecology Department, Med VI Mother and Child Hospital Marrakech, Morocco

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***Corresponding author:** M. Sali
Obstetrics and Gynecology Department, Med VI Mother and Child Hospital Marrakech, Morocco

Abstract **Case Report**

Myasthenia, an autoimmune disease of young women, is due to dysfunction of neuromuscular transmission. The newborn child of a myasthenic mother may suffer from a transient neonatal myasthenic syndrome. Maternal aggravation, or even a myasthenic crisis with respiratory failure, may occur in the first three months post-partum.

Keywords: Myasthenia, pregnancy Summary, Neonatal myasthenia gravis.

INTRODUCTION

Myasthenia gravis is a rare autoimmune disease frequently affecting young women from the second decade onwards, coinciding with the fertile years. The most common course during pregnancy involves a worsening in the first trimester, reaching a peak in the second trimester, followed by remission in the 2nd and 3rd trimesters, with a return to the previous state.

To the previous state [1] Before planning a pregnancy, women with myasthenia must be informed about the management of their treatment during pregnancy, as well as the possibility of exacerbation of the disease and possible fetal risks [1]. Although rare, myasthenia gravis occurs mainly in women of childbearing age, so the association of myasthenia gravis with pregnancy is not exceptional.

Therefore not exceptional. In the post-partum period, myasthenia gravis is frequently aggravated in the mother. Around 12% of newborns born to myasthenic mothers develop transient neonatal myasthenia [1], without any parallelism with the degree of maternal involvement. We report an observation of transient neonatal myasthenia revealing maternal myasthenia.

OBSERVATION

The patient is 24 years old, 3rd gesture, 3rd pare, with a history of a live baby by vaginal delivery, a neonatal death at 6 days of life by vaginal delivery with the onset of a myasthenia gravis crisis in the mother. The patient underwent Caesarean delivery without any abnormalities, with an Apgar score of 10/10. The patient's osserman score was stage I.

At 3 days postpartum, the mother's examination revealed intense asthenia, discreet but undeniable signs of muscular fatigability with progressive dysarthria during conversation, a nasal voice and a poorly expressive facial expression with no swallowing problems - an osserman score of stage II was retained. Focused questioning revealed a very gradual onset of symptoms during pregnancy. An electromyogram with a search for block was carried out on this patient, confirming myasthenia. The mother's evolution was towards rapid decompensation with ptosis and increased bulbar disorders, leading to hospitalization in an intensive care unit.

Aminoglycosides were discontinued as soon as the diagnosis was suspected, as they could be responsible for aggravating the myasthenia [2].

Facial hypomimia, which was the most suggestive warning sign, corrected more slowly over two to three weeks. The mother benefited from anticholinesterase treatment, initiated in response to.

Three months after delivery, she developed swallowing difficulties. A mediastinal CT scan was performed, which revealed thymic hyperplasia, and her antirachial antibodies were very high when she was admitted to intensive care. It was decided to perform a thymectomy. The outcome was favorable.

The neonate, at H3 of age, was admitted to the neonatology unit for whimpering and axial hypotonia. Maternal-fetal infection and fetal distress were ruled out. The dissociated nature of the neurological disorders (refusal to suckle, axial hypotonia, hypomimia, but good contact and normal consciousness) led to a search for a neuromuscular or toxic cause.

Neuromuscular or toxic cause A myasthenia is then evoked and confirmed on the basis of the mother's questioning, the clinical examination and a series of tests (electromyogram, as well as complete biological tests), enabling the diagnosis of transient neonatal myasthenia and early management of a maternal myasthenia crisis in a specialized setting.

In the newborn, spontaneous improvement around day 4 and the absence of signs of severity (effective spontaneous ventilation, absence of false routes) enabled symptomatic treatment with gastric tube feeding for the first few days.

As improvement had already begun when the diagnosis was confirmed, we did not resort to anticholinesterase therapy.

Sucking and tone gradually returned to normal within ten days. Proper bottle-feeding was possible around day 10.

DISCUSSION

This observation reminds us that myasthenia primarily affects women in the second or third decade of life, which overlap with the childbearing years [3]. The course of the disease is unpredictable during pregnancy, with the risk of worsening especially in the third trimester, which was the case for our patient. But myasthenia can be well managed during pregnancy, with relatively safe and effective therapies such as anticholinesterases or even etiological treatment such as thymectomy (outside pregnancy), corticosteroid therapy and immunosuppressants [4]. So women with myasthenia should not be discouraged from conceiving.

However, they should discuss their plans for pregnancy with their neurologist and gynecologist 3 Months in Advance.

From a pathophysiological point of view, two types of antibodies have been described in myasthenic subjects: anti-ACRH and anti-Sm [5].

Transient neonatal myasthenia reveals itself in the first few days of life, often after an interval of a few hours, or up to four days. The classic clinical picture includes feeding difficulties (weak sucking, swallowing disorders), axial and segmental hypotonia, poor mimicry, rapid exhaustion of the cry and hypoventilation [3, 6].

The natural course of the disease is towards regression of signs, which can vary from 15 days to five weeks. This clinical syndrome usually occurs in children of myasthenic mothers already known and treated, and the diagnosis of transient neonatal myasthenia is easily made. The diagnosis is more delicate when there is an intercurrent neonatal disease or when the maternal myasthenia is unknown [7], as is the case in our observation. Indeed, the signs of transient neonatal myasthenia are not specific, and the existence of an infection infection, fetal distress or prematurity [3] can be misleading, especially in the absence of a maternal history. The problem is then to distinguish between these frequent illnesses and to know how to evoke the diagnosis of transient neonatal myasthenia when faced with an isolated and dissociated abnormal neurological examination (refusal to suckle, axial hypotonia, hypomimia, but good contact, normal consciousness). Anticholinesterase testing (Prostigmine) provides rapid diagnostic evidence if it allows a transient regression of symptoms [3]. In our case, confrontation with the mother's findings (clinical monitoring, repeat questioning and history of the observed disorders, electromyogram) provided diagnostic certainty within a short timeframe. The presence of anti-acetylcholine receptor antibodies in the newborn's serum neither confirms nor excludes the diagnosis of transient neonatal myasthenia gravis. Indeed, 10-15% of myasthenic subjects are negative [8], and cases of transient neonatal myasthenia without anti-acetylcholine receptor antibodies have been reported [9]. Moreover, 80% of neonates born to myasthenic mothers have circulating autoantibodies, even though they are free of the condition [10]. Transplacental passage of autoantibodies alone does not explain neonatal disease [7, 10], and it is only in a child who is asymptomatic at birth, and whose mother is known to be myasthenic, that their measurement can be of prognostic interest.

Anticholinesterase agents can be used in full-term neonates [4], and have also been used in premature infants without major side effects [3]. Other treatments are occasionally reported: exchange transfusions [4], high-dose immunoglobulin infusions [12]. Only severe forms, with fetal repercussions, are detectable in utero. Ultrasound can reveal excess amniotic fluid, reduced fetal movement, absence of swallowing and, in the most severe forms, even fetal immobility syndrome [4, 7, 13]. Here again, the absence of known myasthenia in the mother should not rule out the diagnosis [7, 13]. At birth, these are always severe forms in which complications of fetal immobility syndrome (arthrogryposis) are added to the usual clinical picture [4, 7, 12, 13].

Antibody studies in newborns with transient neonatal myasthenia reveal the existence of IgG of different idiotypes from those of their mothers, indicating the synthesis of anti-acetylcholine receptor antibodies in the newborn [10, 15], possibly secondary to transplacental passage of maternal immunocytes [16].

The immaturity of fetal acetylcholine receptors could also explain their insensitivity to antibodies [17].

Above all, alpha-fetoprotein is thought to have a protective effect, inhibiting the binding of antibodies to acetylcholine receptors [18]. The post-partum decreases in of circulating alpha-fetoprotein in both mother and child could explain both the transient neonatal myasthenia and the worsening or, as in our observation, onset of maternal symptoms.

CONCLUSION

Myasthenia gravis has little effect on the course of pregnancy. Pregnancy, however, can alter the stability of this pathology, requiring redoubled attention and full cooperation from a multidisciplinary medical team. Regular neurological monitoring during pregnancy and post-partum should be instituted to detect relapses at an early stage, and to institute specific treatments to curb them. Anesthesia consultation should be carried out as early as possible. Scheduling the delivery in agreement with the obstetric team helps to limit the risks, with vaginal delivery remaining the preferred option. Analgesia is highly beneficial, and locoregional analgesia using low concentrations of local anesthetic is recommended. Finally, close monitoring of the post-partum period can detect decompensation. Transient neonatal myasthenia does not correlate with the severity of the maternal disease, which it may reveal [4].

In the neonatal period, management requires rapid diagnostic confirmation, based above all on rigorous analysis of clinical semiology, anticholinesterase testing, and maternal clinical examination and electromyogram. Thus, only good obstetric-pediatric collaboration can prevent diagnostic errors and enable early, appropriate care of mother and child.

REFERENCES

1. Lucot, J. P., Dufour, P., Vinatier, D., Tordjeman, N., Durant-Reville, M., & Puech, F. (1996). Myasthénie et grossesse: A propos de 2 observations. *Journal de gynécologie obstétrique et biologie de la reproduction*, *25*(2), 179-185.
2. Dunand, M., Lalive, P. H., Vokatch, N., & Kuntzer, T. (2007). Myasthéniegrave, traitements. *Rev Med Suisse*, *3*, 1185-1190.
3. Ferrero, S., Pretta, S., Nicoletti, A., Petrera, P., & Ragni, N. (2005). Myasthenia gravis: management issues during pregnancy. *European Journal of Obstetrics & Gynecology and Reproductive Biology*, *121*(2), 129-138.
4. Danziger, N., & Alamowitch, S. (2019). Neurologie. 13e éd. actualisée. Paris: Med-Line éditions; 2019. (Le référentiel).
5. Eymard, B., Vernet-der Garabedian, B., Berrih-Aknin, S., Pannier, C., Bach, J. F., & Morel, E. (1991). Anti-acetylcholine receptor antibodies in neonatal myasthenia gravis: heterogeneity and pathogenic significance. *Journal of autoimmunity*, *4*(2), 185-195.
6. Váradi, V., Nagy, A., György, I., Machay, T., & Papp, Z. (1995). Transient neonatal myasthenia gravis. *Orvosi Hetilap*, *136*(8), 401-405.
7. Riemersma, S., Vincent, A., Beeson, D., Newland, C., Hawke, S., Vernet-der Garabedian, B., ... & Newsom-Davis, J. (1996). Association of arthrogryposis multiplex congenita with maternal antibodies inhibiting fetal acetylcholine receptor function. *The Journal of clinical investigation*, *98*(10), 2358-2363.
8. Vincent, A., Li, Z., Hart, A., Barrett-Jolley, R., Yamamoto, T., Burges, J., ... & Newsom-Davis, J. (1993). Seronegative Myasthenia Gravis: Evidence for Plasma Factor (s) Interfering with Acetylcholine Receptor Function a. *Annals of the New York Academy of Sciences*, *681*(1), 529-538.
9. Orradre, C. O., Serrano, E. N., Moises, V. R., Pison, J. L., Traid, A. V., & Tello, A. M. (1996). Seronegative transient neonatal myasthenia: report of a case. *Anales espanoles de pediatria*, *45*(6), 651-652.
10. Eymard, B., Morel, E., Dulac, O., Moutard-Codou, M. L., Jeannot, E., Harpey, J. P., ... & Bach, J. F. (1989). Myasthenia and pregnancy: a clinical and immunologic study of 42 cases (21 neonatal myasthenia cases). *Revue neurologique*, *145*(10), 696-701.
11. Lefvert, A. K., & Per Olof Osterman. (1983). Newborn infants to myasthenic mothers: AL clinical study and an investigation of acetylcholine receptor antibodiesin 17 children. *Neurology*, *33*(2), 133-138.
12. Bassan, H., Muhlbaur, B., Tomer, A., & Spirer, Z. (1998). High-dose intravenous immunoglobulin in transient neonatal myasthenia gravis. *Pediatric neurology*, *18*(2), 181-183.
13. Verspyck, E., Mandelbrot, L., Dommergues, M., Huon, C., Woimant, F., Baumann, C., & Garabedian, B. V. D. (1993). Myasthenia gravis with polyhydramnios in the fetus of an asymptomatic mother. *Prenatal diagnosis*, *13*(6), 539-542.
14. Eymard, B., Vernet-der Garabedian, B., Berrih-Aknin, S., Pannier, C., Bach, J. F., & Morel, E. (1991). Anti-acetylcholine receptor antibodies in neonatal myasthenia gravis: heterogeneity and pathogenic significance. *Journal of autoimmunity*, *4*(2), 185-195.
15. Pilkington, C., Lefvert, A. K., & Rook, G. A. W. (1995). Neonatal myasthenia gravis and the role of agalactosyl IgG. *Autoimmunity*, *21*(2), 131-135.
16. Morel, E., Eymard, B., Garabedian, B. V. D., Pannier, C., Dulac, O., & Bach, J. F. (1988). Neonatal myasthenia gravis: a new clinical and immunologic appraisal on 30 cases. *Neurology*, *38*(1), 138-142.
17. Vernet-der Garabedian, B., Lacokova, M., Eymard, B., Morel, E., Faltin, M., Zajac, J., ... & Bach, J. F. (1994). Association of neonatal myasthenia gravis with antibodies against the fetal acetylcholine receptor. *The Journal of clinical investigation*, *94*(2), 555-559.
18. Brenner, T., Abramsky, O., & Lisak, R. P. (1981). Influence of alpha-fetoprotein on the in vitro and in vivo immune response to acetylcholine receptor. *Annals of the New York Academy of Sciences*, *377*, 208-221.