CASE REPORT

Spontaneous Recovery of Neuromyelitis Optica Spectrum Disorder during Pregnancy

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ABSTRACT

Neuromyelitis optica spectrum disorder (NMO-SD) is an autoimmune inflammatory disorder associated with the anti-aquaporin-4 (AQP4) antibody. Over 90% of NMO-SD patients have poor prognosis, and pregnancy is a disease-worsening factor. The authors report the findings in a case of NMO-SD that recovered spontaneously during pregnancy. A 28-years-old woman developed optic neuritis (ON) of the right eye in the 14th week of pregnancy. Her best-correlated visual acuity (BCVA) was 0.02 OD, the critical flicker frequency (CFF) was not measurable, and the mean deviation (MD) of the Humphrey perimetry 30-2 was -42 dB. Magnetic resonance imaging (MRI; T2 short-tau inversion recovery [STIR]) showed high-intensity right optic nerve. The patient was diagnosed with NMO-SD by the detection of seropositive anti-AQP4 antibody in her serum. Corticosteroid pulse therapy was considered, but the patient declined any treatment. Two weeks later, her BCVA improved to 1.2 (OD). Nine weeks later, the MD of Humphrey perimetry 30-2 improved to -1.2 dB and the CFF was 39.8 Hz. There have been no signs of recurrence for 6 months. Spontaneously recovered cases of NMO-SD during pregnancy are very rare. Although the factors leading to good outcomes have not been identified, this case suggests that the immunological condition at the early stage of the pregnancy may have different from that at later stages. Further studies are needed to identify the relationship between pregnancy and NMO-SD.

Keywords: Neuromyelitis optica, optic neuritis, pregnancy, spontaneous recovery

INTRODUCTION

Neuromyelitis optica (NMO) is a recurrent demyelinating autoimmune disease that causes severe damage to the optic nerve and the spinal cord. NMO recently gained its place among the autoimmune diseases, although the first detailed case of NMO was reported by Devic as early as 1894.¹ NMO was initially misdiagnosed as multiple sclerosis (MS) because there was no biological marker that could distinguish these two diseases. However, after the discovery of the anti-aquaporin-4 (AQP4) antibody, the understanding of the pathogenesis of NMO has improved considerably.²

AQP4 is a water channel of the astrocytes in the central nervous system, and the antibody against this channel plays a key role in the pathology of NMO. The concept of NMO was broadened by the discovery of this antibody, and now optic neuritis with seropositive anti-AQP4 antibody without myelitis is classified as NMO spectrum disorder (NMO-SD).³

The clinical course of NMO-SD is recurrent and severe, and it is mostly resistant to corticosteroid therapy. Thus, patients with NMO-SD require plasma exchange or other immune suppressive therapy.⁴ Pregnancy is reported to be a NMO-SD-worsening factor, and the relapse rate of NMO rises after birth.⁵

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FIGURE 1 Initial findings of (a) Humphrey perimetry 30-2 and (b) MRI (T2 STIR). The MD of left eye was -1.66 dB and the MD of right eye was -24.62 dB (a). MRI (T2 STIR) of brain and optic nerve shows high-intensity signals in the right optic nerve (b).

We report a case of NMO diagnosed during the early stage of the pregnancy that had a spontaneous recovery without any treatment.

CASE REPORT

The patient was a 28-year-old woman who presented with an acute visual decrease of her right eye and pain on ocular movements. The patient was in her 14th week of pregnancy at her first visit to our clinic. Her best-corrected visual acuity (BCVA) was 0.02 OD, the relative afferent pupillary defect was positive in the right eye, the critical flicker frequency (CFF) was not measurable, and the mean deviation (MD) of Humphrey perimetry 30-2 was -42 dB. Magnetic resonance imaging (MRI; T2 short-tau inversion recovery [STIR]) examination showed high-intensity signals in the right optic nerve, and there were no signs of a brain lesion (Figure 1). MRI of the spinal cord was not conducted because there were no abnormalities in the neurological examinations. Haematological examinations showed no signs of

autoimmune diseases. An enzyme-linked immunosorbent assay (ELISA) was conducted to determine whether anti-AQP4 antibody was present, and the results showed that the patient was seropositive for anti-AQP4 antibody.

The patient was diagnosed with NMO-SD by the presence of optic neuritis and seropositive anti-AQP4 antibody. Corticosteroid pulse therapy was considered, but the patient declined any treatment because she was in the first trimester of her pregnancy. Two weeks later, her BCVA improved to 1.2 OD and the CFF was 17.8 Hz. Nine weeks later, the MD of Humphrey 30-2 improved to -1.2 dB and the CFF also increased to 39.8 Hz (Figure 2). The patient delivered at 39 weeks without any complications. There have been no signs of recurrence for at least 6 months.

DISCUSSION

Pregnancy is reported to have a negative influence on the manifestation of NMO-SD.⁵ There are few case reports of NMO-SD with good outcomes, e.g., a case

FIGURE 2 Humphrey perimetry 30-2 6 months after the first visit. For the Humphrey perimetry 30-2, the MD of the left eye was -2.13 dB and MD of the right eye was -1.34 dB.

at 8-week pregnancy that had a complete recovery after treatment with corticosteroid,⁶ and a French multicentre cohort study reported that 11 patients out of 175 showed good outcome after treatment with corticosteroid or other immunosuppressive treatment.⁴ However, most cases of the NMO-SD during pregnancy had an unfavourable course, with severe vision reduction and residual movement impairments.⁴ At the present time, there is no case report that demonstrated the recovery of NMO-SD without treatment during pregnancy. The annual relapse rate of NMO-SD rises during the first and second trimesters after birth.⁵

Immunological changes during pregnancy and the disease-worsening factors remain open to question. The results of recent studies provided some clues on the role of immunological factors. Immunity is composed of cell-mediated immunity and humoral immunity, and the Th2 (T-helper cell type 2) cytokines related to humoral immunity predominate during pregnancy.⁷ A shift away from cell-mediated immunity, i.e., Th1-mediated immunity, toward humoral immunity, i.e., Th2-mediated immunity, in the early stage of pregnancy plays an important role in maintaining normal pregnancy. Cell-mediated autoimmune diseases such as multiple sclerosis (MS) and rheumatic arthritis (RA) are known to be ameliorated during pregnancy, but humoral autoimmune diseases such as NMO are exacerbated because of this shift.⁸ The degree of this shift is reported to differ between individuals.⁹

Our hypothesis for our case is that there was an incomplete shift from Th1 to Th2, which could have prevented disease progression. This incomplete shift may also have prevented relapses after birth. However, this hypothesis cannot be tested because

we did not measure the cytokine levels in our patient. Clinically, the findings in our case suggest that NMO-SD at the early stage of pregnancy has a possibility of having a spontaneous recovery without treatment. Corticosteroid and other immunosuppressive medications always have chances of side effects that could affect the embryo, and the choice of observation alone is an option that can avoid these risks. The treatment options for patients with NMO-SD during pregnancy should be decided after it is well discussed with patient.

The limitation of this is only one case, and the lack of immunological examination limits our prediction for future cases. Further investigations are needed to identify the factors that lead to good outcome NMO-SD during pregnancy.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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