Challenging Management of Neuromyelitis Optica during Pregnancy

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Introduction

Neuromyelitis optica (NMO) is a less common CNS demyelinating disease presenting with relapsing course, but clinical and laboratory features distinctly different from typical multiple sclerosis (MS). NMO relapses can be very severe and the prognosis devastating. NMO is more common in females but little information is available regarding clinical course and management of NMO during pregnancy.

Case

22yo Asian/Native American female diagnosed with NMO at age 19 per classic clinical and MRI presentation; negative NMO-IgG-antibodies in CSF and serum. Developed co-morbidity of insulin-dependent DM Type 2 at age 20.


Became unexpectedly pregnant while on AZA and with untreated IDDM (HbA1c 12.8). Only spotty early maternal care.

During 2nd and 3rd trimester repeatedly severe NMO-relapses requiring multiple iv steroid treatments (IVMP), plasmapheresis (PLEX), and iv immune globulin (IVIG). Due to severity of disease, she was delivered at week 32 for escalation of immune therapy (rituximab).

Post-partum renewed disease activity (contralateral vision impairment, worsened LUE weakness) 2 months after rituximab 2x 1000mg while fully B cell-depleted. Thus far, incomplete recovery after 5 day IVMP and 7x PLEX. Recently started on add-on AZA.

Discussion

This case illustrates that NMO can be very active and management challenging during pregnancy. A young woman lost significant function (R eye vision loss, weakness BUE) in setting of poorly controlled disease activity during pregnancy despite aggressive therapy with repeated IVMP, PLEX and IVIG -and- post-delivery her disease reoccurred after initiation of rituximab despite complete B cell depletion.

- Does typical clinical course during and after pregnancy differ in NMO versus MS?
- What else could have been done to reduce NMO-disease activity during pregnancy?
- Would have outcome been better if she had received cytoxan instead of rituximab?
- How to manage patients with NMO-disease activity despite rituximab?

Baby boy Z.

- Born at gestational age 32 weeks 0 days by elective cesarean. Weight 1600g (26th percentile), length 39.5 cm (8th percentile), APGAR 6/7, acrocyanotic, quiet, floppy
- Respiratory distress syndrome, intubated x 1 day, weaned to room air at 1 week
- Apnea of prematurity x 3 weeks
- Hyperbilirubinemia, phototherapy x 3 days
- Possible sepsis at age 2 weeks
- Aspiration at age 6 weeks
- Discharge at age 8 weeks, weight 3600g (48th)

Pregnancy

- Acute onset paroxysm and painful paresthesia at week 20, 3 day IVMP.
- Rapid deterioration and complicated clinical course with significant ups & downs; at times RUE / BLE paresis, non-ambulatory, R eye vision loss with interim response to repeated IVMP and PLEX; then restarted AZA while pregnant.
- At ~ week 30, again acute worsening with progressive tetraplegia and R eye complete vision loss, non-responsive to repeat IVMP, PLEX and IVIG. Elective cesarean at week 32 for escalation immune therapy.
- In the end, sustained R eye vision loss (light perception only), weakness BUE interfering with activities and multiple drugs for paresthesia.