Postpartum Optic Neuropathy: Think of Myelin Oligodendrocyte Glycoprotein Immunoglobulin G-Associated Optic Neuritis - Report of Two Cases

Dear Sir,

Myelin oligodendrocyte (MOG) immunoglobulin G (IgG)-associated disorder (MOGAD) has a broad range of clinical manifestations that include optic neuritis (ON), transverse myelitis (TM), acute disseminated encephalomyelitis (ADEM), and brainstem encephalitis.[1] There are various causes of postpartum visual disturbances with preeclampsia, eclampsia causing posterior reversible leukoencephalopathy to be more common. However, the acute first attack of central nervous system demyelination in the postpartum period is relatively uncommon. Hereby, we report two patients with bilateral optic neuritis who had the first attack in the postpartum period and were seropositive for MOG-IgG. This occurrence of ON due to MOG-IgG in the postpartum period has not been commonly described.

Case 1

A 22-year-old lady who delivered a healthy baby by normal vaginal delivery 15 days back presented to the hospital with a history of painful progressive loss of vision of 1-week duration. The diminution of vision was initially noticed in the left eye followed by the right eye within 1 day. There was no redness of eyes or discharge from the eyes. There was no history of headache, vomiting, fever, and joint pains. There was no previous history of visual disturbances, preeclampsia during pregnancy. Higher mental functions were normal. The fundus examination showed disc edema in both eyes with the normal macula. Visual acuity was the perception of light in both the eyes. Pupillary light reflex was sluggish in both eyes. The rest of the neurological examination was normal. Complete hemogram including erythrocyte sedimentation rate, renal, liver, and thyroid function tests, serum electrolytes, and serum angiotensin-converting enzyme were normal. Antinuclear antibody profile and anti-nuclear cytoplasmic antibodies were negative. Visual-evoked potentials showed absent P100 waveform in both eyes. Brain and spine with contrast magnetic resonance imaging (MRI) showed mild T2 and fluid-attenuated inversion recovery (FLAIR) hyperintensity involving the intraorbital optic nerve Figure 1]. No parenchymal or cord lesions were noted. Cerebrospinal fluid (CSF) analysis showed normal CSF opening pressure, cell count (2 cells- lymphocytes), protein (27 mg/dL), and glucose (63 mg/dL). Serum and CSF **anti-MOG** antibodies (qualitative assessed using cell-based assay) were **strongly positive**. Serum,

CSF aquaporin-4 antibodies and CSF oligoclonal body were negative. She was treated with the intravenous methylprednisolone (IVMP) (1 g per day for 5 days) followed by large volume plasmapheresis (LVPP) (5 cycles) as there was no improvement with steroids. There was mild improvement in vision in both eyes (right eye- counting finger 4 meters and left eye- counting finger 2 meters). She was discharged with oral steroids. Repeat serum **anti-MOG** antibodies were negative.

Case 2

A 21-year-old lady who delivered a healthy baby by normal vaginal delivery 2 months back presented to the hospital with a history of painful progressive loss of vision of 11-days duration. The diminution of vision was initially noticed in the right eye followed by the left eye within 3 days. There was no redness of eyes or discharge from the eyes. There was no previous history of visual disturbances, preeclampsia during pregnancy. Higher mental functions were normal. The fundus examination was normal. Visual acuity was close to face in the right eye and perception of light in the left eye. Pupillary light reflex was sluggish in both the eyes. The rest of the neurological examination was normal. Complete hemogram including erythrocyte sedimentation rate, renal, liver, and thyroid function tests, serum electrolytes, and serum angiotensin-converting enzyme were normal. Antinuclear antibody profile and anti-nuclear cytoplasmic antibodies were negative. Visual-evoked potentials showed absent P100 waveform in both eyes. Brain and spine with contrast magnetic resonance imaging (MRI) showed T2 hyperintensity involving the intraorbital optic nerve [Figure 2]. No parenchymal or cord lesions were noted. Cerebrospinal fluid (CSF) analysis showed normal CSF opening pressure, cell count (1 cell- lymphocyte), protein (16 mg/dL), and glucose (58 mg/dL). Serum and CSF anti-MOG antibodies (assessed using cell-based assay) were strongly positive. Serum, CSF aquaporin-4 antibodies and CSF oligoclonal body were negative. She was treated with the IVMP (1 gram per day for 5 days) and LVPP (5 cycles) as there was no improvement with steroids. There was a significant improvement in vision in both eyes (right eye- 6/6 and left eye- counting finger 3 meters). She was discharged with oral steroids.

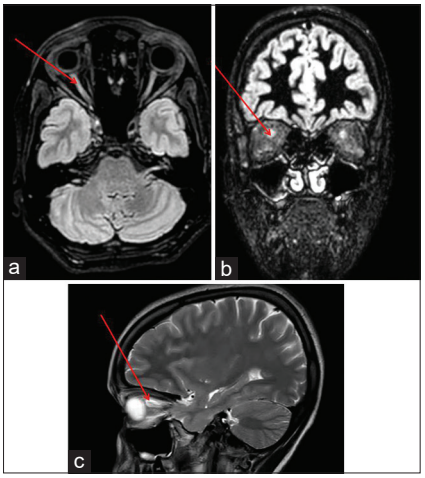

Figure 1: Brain magnetic resonance imaging of case 1 showing (a) hyperintensity in optic nerve (red arrow) axial fluid-attenuated inversion recovery (FLAIR) image; (b) hyperintensity in optic nerve (red arrow) coronal double inversion recovery (DIR) image; (c) hyperintensity in optic nerve (red arrow) sagittal T2 image

Discussion

Acute vision loss in the postpartum period has several reported etiologies that include severe preeclampsia/eclampsia (in which vision loss could be due to exudative retinal detachment, hypertensive retinopathy, and cortical blindness), posterior reversible leukoencephalopathy syndrome, pituitary apoplexy (thunderclap headache, vomiting, visual loss), posterior ischemic optic neuropathy (PION), anterior ischemic optic neuropathy (AION), cortical venous sinus thrombosis, and central serous retinopathy.[2-6] Sudden hypotension complicated with anemia can lead to acute bilateral simultaneous PION. The fundus examination will be normal in PION. Nonarteritic AION presents with a sudden visual loss with segmental disc edema on fundus examination. Bilateral ON due to MOGAD is an uncommon cause of visual loss in the postpartum period.

MOG is a central nervous system protein expressed on the surface of the oligodendrocytes. Antibodies against MOG have been implicated in the demyelination, which has been exemplified in the animal models of demyelination. Males and females are affected in equal proportion in MOGAD. ON is the commonest presentation. An UK study by Jurynczyk M *et al*., (2017) showed that ON were the presenting symptoms in 55% of patients with MOGAD.[7] A multicenter epidemiologic survey of ON between 2015 and 2018 from Japan found that 10% were positive for MOG-IgG (out of 531 serum samples).[8] ON in MOGAD are usually recurrent (50% of cases), bilateral, and associated with optic disc edema (86% of cases) even peripapillary hemorrhage. ON due to MOGAD can be confused for papilloedema and nonarteritic anterior ischemic optic neuropathy due to frequent occurrence of disc edema.

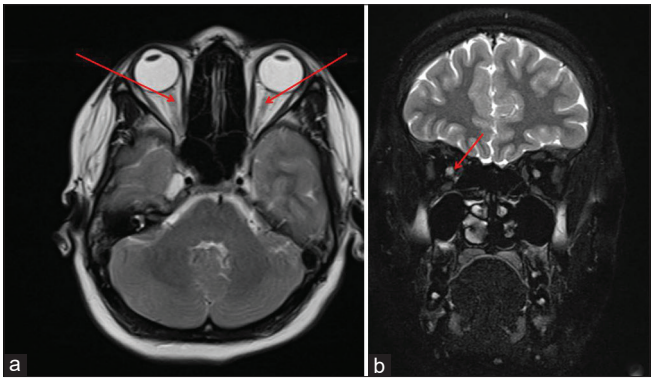

Figure 2: Brain magnetic resonance imaging of case 2 showing (a) hyperintensity in optic nerve (red arrow) axial T2 image; (b) (a) hyperintensity in optic nerve (red arrow) coronal T2 image

ON in MOGAD have a severe visual loss at presentation but show remarkable recovery with steroids, and can have relapse once the steroid is tapered off or stopped.[9] ON affects a long segment of the intraorbital portion of the optic nerve with contrast enhancement of optic nerve sheath and surrounding orbital fat tissue. The acute management of MOG-IgG positive ON is 1 g per day of IVMP for 3–5 days. Plasma exchange is also recommended as MOGAD is an antibody-mediated disease if the visual loss is severe and nonresponsive to IVMP.

There is a relation between the state of pregnancy and symptoms of CNS demyelinating disorders. It may ameliorate, deteriorate, or show no changes at all. Multiple sclerosis patients have a lower relapse rate during pregnancy, with an increase in the relapse rate during the postpartum period. There is exacerbation in the attacks of neuromyelitis optica during pregnancy and recurrence during the postpartum period. A multicenter study on 50 patients with MOGAD had 3 patients in whom MOGAD occurred during the postpartum period with one patient having a denovo ON attack during the early postpartum period. The immunological changes during pregnancy and after delivery may trigger an attack or cause disease induction.[10]

Conclusion

MOGAD is a recently described CNS demyelinating disease and ON is the most common presentation of MOGAD. ON is recurrent, bilateral with severe visual loss during an acute attack. An acute first attack of ON due to MOGAD in the postpartum state is uncommon. The treating physician and obstetrician should be aware of this recent described CNS demyelinating disease as a cause of acute visual loss in the postpartum period.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 18-Apr-2020 **Revised:** 03-May-2020 **Accepted:** 11-May-2020
Published: 22-Jul-2020

DOI: 10.4103/aian.AIAN_317_20