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Case Report

A 66-Year-Old Woman Presented with Sudden Onset Eye Pain and Loss of Vision: Do Not Miss MOGAD

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Abstract

A 66 -year- old woman presented to ED with sudden onset eye pain and unilateral loss of vision. She had considerable vascular risk factors including age, hypertension, diabetes mellitus and hypercholesterolemia. She was referred to neurology service with possible arteritic anterior ischemic optic neuropathy (AION). Given her age, painful visual loss and raised ESR, she was treated with steroids for possible giant cell arteritis (GCA). Because of her psychiatric history and poorly controlled diabetes mellitus, steroids were weaned quickly. 6 months later, she had a similar presentation affecting the other eye. MRI head showed disease progression with positive myelin oligodendrocyte glycoprotein (MOG) antibody in serum. MOG associated disorder (MOG-AD) is a steroid responsive inflammatory demyelinating disease but needs long term immunosuppression plan and monitoring of serum antibody.

***Keywords:** MOG-AD, Optic neuropathy, Corticosteroid*

Introduction

MOG-AD is a recognized inflammatory demyelinating disease that is distinct from multiple sclerosis and acuaporin-4 neuromyelitis optica (AQP-4 NMO). It has a wide variable clinical presentation including classical optic neuritis, myelitis, Acute Disseminated Encephalomyelitis (ADEM) and cortical encephalitis. The diagnosis depends on determination of specific and sensitive assay of antibody. It can have a monophasic course or a relapsing course. Long term use of steroid and disappearance of the antibody likely reduce the risk of relapse.

Case Presentation

A 66-year-old lady presented to our emergency department with a sudden onset left orbital sharp and constant pain for 7 days. The pain was localized, worsened with horizontal eye movements, and improved with naproxen. Additionally, she noticed gradual visual impairment of the same side over the subsequent days. There was accompanied jaw claudication. There were no other neurological or systemic manifestations.

Her medical history includes hypertension, type-2 diabetes mellitus, hypercholesterolemia, schizophrenia and essential tremors.

Initially, she was seen by her general practitioner who thought she had conjunctivitis. Despite treating her with Chloramphenicol, the pain and visual impairment had progressed which warranted an ophthalmologic referral. Ophthalmic examination revealed visual acuity of counting fingers on the left eye and 6/24 on the right eye. There was left optic disc swelling and no relative afferent pupillary defect (RAPD) bilaterally. Visual fields and color vision were hard to illicit due to poor vision on the left eye. At this point, she was given a clinical diagnosis of arteritic ischemic optic neuropathy (AION) and referred to neurology.

When seen in the neurology urgent clinic, she had no headache, temporal artery abnormality or other focal signs. The ESR was 54 mm/h.

In a 66-year-old person a presentation of rapidly progressive painful visual loss, jaw claudication and raised ESR; giant cell arteritis should be considered and treated until proven otherwise. She has vascular risk factors to predispose to NAION as the second possible diagnosis. Inflammatory optic neuropathy felt to be less likely with no RAPD.

Intravenous methylprednisolone course for 3 days was given followed by oral prednisolone.

Three-days methylprednisolone led to minor improvement of her vision. She started seeing shapes clearly on left eye but was unable to count fingers at 30 cm. Parenteral steroids was followed by oral prednisolone 60 mg daily with 10 mg weekly wean to stop after 6 weeks. During the follow-up in the clinic 4 months later, the left eye visual acuity improved to 6/15-2 and the left optic disc became pale. Other neurological examination was normal at this stage. As a result, she was referred to formal ophthalmological assessment with OCT and repeat brain imaging before considering restarting steroids in the view of significant mental health history and diabetes.

Formal ophthalmic assessment showed left visual acuity of 6/60, pale left optic disc and colour vision using Ishihara plate was 2/15. Optic Coherence Tomography (OCT) revealed thinning of the retinal nerve fiber layer (RNFL) from previous optic neuritis. Two days prior repeating the scheduled MRI, she developed painful vision loss of the right eye. Unfortunately, the patient did-not seek immediate medical advice at the time but, she rather preferred to wait for the MRI. At this stage, the examination revealed that she was unable to count fingers accurately by the right eye, loss of color perception and RAPD. The left eye visual acuity was still 6/60. Blind spots were enlarged bilaterally, and there was bilateral pale optic disc that is more established on the left side.

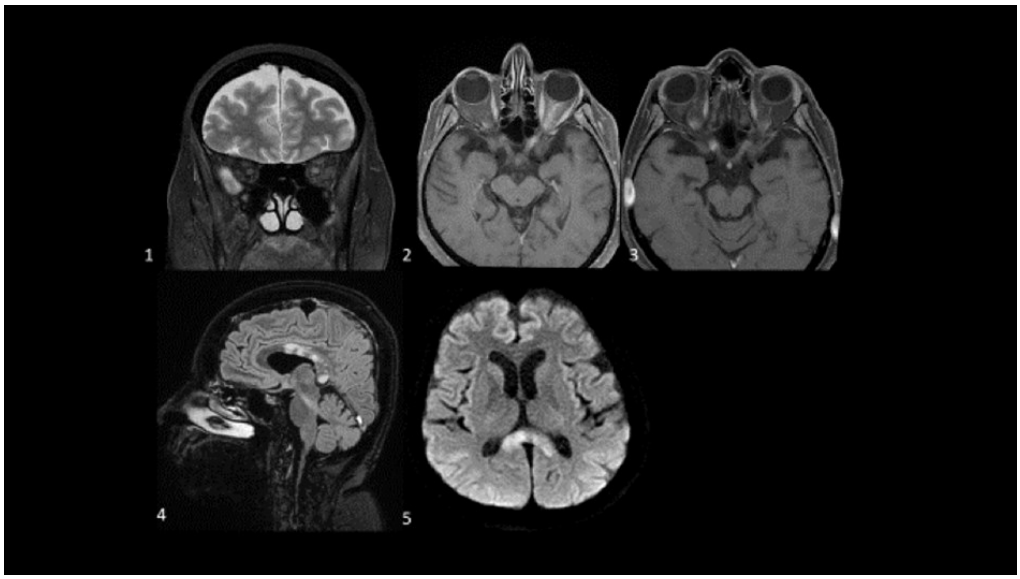

Figure 1: *T2 (1) and fat saturated T1 post contrast (2) imaging from presentation showing T2 hyperintensity and contrast enhancement of a long segment of the intraorbital and canalicular left optic nerve. A follow up study 5 months later shows enhancement of the distal canalicular and prechiasmatic right optic nerve on fat saturated T1 post contrast imaging (3) with hyperintense lesions on T2 FLAIR (4) within the body and splenium of the corpus callosum that exhibit diffusion restriction (5).*

Investigations

In the initial presentation, computed tomography (CT) of the brain and CT angiogram of carotid and intracerebral arteries were normal apart from increased caliber and tortuous left optic nerve.

Brain and orbit magnetic resonance imaging (MRI) with contrast confirmed left optic nerve swelling with contrast enhancement involving infraorbital, intracanalicular and post chiasmatic segments of the left optic nerve. Within the brain, there are a few tiny T2/FLAIR hyperintense foci white matter abnormality which looked entirely nonspecific.

As these findings were suggestive of active inflammatory left optic neuritis, lumbar puncture was performed. CSF analysis showed normal protein, negative oligoclonal bands and normal cells. Aquaporin-4 antibody was negative while myelin oligodendrocyte glycoprotein (MOG) antibody could not be analyzed due to insufficient sample. Serum ANA, ENA, ANCA, DsDNA, Cardiolipin IgG and IgM, C3, C4, HIV, HTLV, Hep B core Ab, Hep C Ab, Syphilis, Borrelia Ab, Aquaporin 4 antibody were negative while MOG Ab was positive.

As she presented again with symptoms of optic neuritis, We repeated MRI head and orbit with contrast along with MRI whole spine. These revealed a significant disease progression. There was bilateral orbital, callosal, brainstem and cranial nerve disease which were keeping with an inflammatory demyelinating disease and are supportive of MOG antibody disease.

Treatment

She was admitted after the second presentation and started on intravenous methylprednisolone for 3 days followed by oral prednisolone 60 mg with a slow weaning plan. Moreover, she had given five plasma exchange sessions. Mycophenolate mofetil was started with a plan to closely monitor her in the clinic.

Outcome and follow up

There was no noticeable improvement at the early stage after admission. She was seen 3 months later with mild improvement in her vision and no further relapses.

Discussion

MOG-AD is a neuro-inflammatory condition that preferentially affect the optic nerve but also the spinal cord and brain. MOG is a protein that is located on the surface of myelin sheaths of the central nervous system[1].

The incidence and prevalence of MOGAD are largely unknown, although studies in Europe suggest the incidence of MOGAD is between 1.6 and 3.4 per 1,000,000 person-years[2]. Children account for up to half of reported cases. In one report, the incidence of MOGAD was higher in children compared with adults (3.0 versus 1.3 per million person-years, respectively)[3]. The median age of MOGAD onset is 20 to 30 years[4].

While none of the clinical features of MOGAD are disease-specific, some are highly characteristic. These include acute attacks of unilateral or bilateral optic neuritis resulting in severe visual loss; acute disseminated encephalomyelitis (ADEM); transverse myelitis which characteristically involves long segment of the spinal cord and has a predilection to affect conus medullaris[5].

Other central nervous system (CNS) involvement may occur, including the clinical syndrome of neuromyelitis optica spectrum disorder (NMOSD) without aquaporin-4-immunoglobulin G (AQP4-IgG) detected[6]. Patients with MOGAD may also present with unilateral cerebral cortical encephalitis resulting in seizures, headache, aphasia, stroke like episodes and fever[7]. The brainstem can also be involved[8].

Clinical attacks usually develop over days and may plateau with variable recovery over weeks to months. The neurological manifestations may be preceded by an infectious illness or vaccination[9].

MOGAD differs from MS and AQP4-IgG positive NMOSD in that a large proportion of patients (40 to 50%) have a single attack without recurrence–(a monophasic course). In a series of 366 patients, 56% of adults and 53% of children had relapsed after a median follow-up of 2.5 to 3 years[5].

Most relapses occur in the early months after the onset. The risk of relapse is lower in patients treated with prolonged steroids course, compared with patients treated for shorter periods of time. Moreover, the disappearance of MOG is typically associated with cessation of relapses[10].

Proposed diagnostic criteria from a single center for MOG antibody-associated disease require the following:[11]

Serum positivity for MOG-IgG by cell-based assay

A clinical presentation consistent with any of the following central nervous system syndromes:

- ADEM
- Optic neuritis
- Transverse myelitis
- Brain or brainstem demyelinating syndrome
- Any combination of these
- Exclusion of an alternative diagnosis.

Acute relapses are usually treated immediately, and the typical regimen comprises a course of 3–7 days of high-dose (0.5 –1 g) intravenous or oral methylprednisolone followed by oral prednisolone. Patients with severe attacks or who respond poorly to corticosteroids should be treated early with plasma exchange (or intravenous immunoglobulin) with the aim of improving recovery[12].

The relapse risk is highest in the early months after disease onset, and follow-on prednisolone cover seems to reduce the relapse rate. The recommended clinical practice is to maintain those patients on a low dose oral prednisolone (e.g., 10 mg daily) for 6 months after the initial attack. Disappearance of MOG antibody seems to indicate remission. This should be retested at 6 months. If it is negative-corticosteroids can be stopped[12].

However, it is reasonable to consider treating patients with persistent antibodies for up to 12 months. It is the clinical practice switching to a steroid-sparing agent if a longer duration of immunosuppression is required[12].

Conclusion

It is crucial to consider MOG-AD as a possible cause of optic neuropathy despite the age of the patient. Patients are highly responsive to steroids, but vulnerable to relapse on steroid reduction and cessation.

Conflict of Interest

The authors have no conflicts of interest to declare.

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