

A Case of HLA-B27-Associated Optic Neuritis

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Abstract

We report the case of a young woman who presented with chronic relapsing inflammatory optic neuritis (CRION) in association with HLA-B27 and silent brain lesions. The attacks were refractory to steroids but no further attacks occurred on treatment with mycophenolate mofetil. This is one of the few HLA-B27-associated optic neuritis cases reported in the literature, emphasizing the relation of HLA-B27 and inflammation of the brain and optic nerve, as well as the role mycophenolate mofetil might offer in stabilizing the disease.

Keywords: Optic neuritis • HLA-B27 • Demyelinating • Vasculitis

Abbreviations

(IVMP) Intravenous Methyl Prednisolone; (RNFL) Retinal Nerve Fiber Layer; (VEP) Visual Evoked Potentials; (MRI) Magnetic Resonance Imaging; (MRA) Magnetic Resonance Angiography; (CSF) Cerebrospinal Fluid; (CRION) Chronic Inflammatory Relapsing Optic Neuropathy; (MMF) Mycophenolate Mofetil; (ON) Optic Neuritis; (NMO) Neuromyelitis Optica; (MS) Multiple Sclerosis; (AS) Ankylosing Spondylitis

Introduction

Chronic relapsing inflammatory optic neuropathy (CRION) is an autoimmune disease of the optic nerve that can present with pain and vision loss, and is characterized by its dependency on immunosuppressive therapy, with relapses upon withdrawal of treatment [1]. Rare cases of optic nerve and brain inflammation have been associated with HLA-B27, in patients who did not fit overt criteria for typical neuro-demyelinating disease such as multiple sclerosis (MS) and neuromyelitis optica (NMO). We add to the literature by describing a case of HLA-B27-associated optic neuritis (ON) and silent brain lesions, who achieved disease stabilization on mycophenolate mofetil (MMF).

Case Presentation

A 37 year old lady, ex-smoker with migraine disorder, presented for isolated painful blurry vision in her right eye, without any other focal neurological deficit. Her exam consisted of decreased vision in the right eye (20/200) and normal vision in the left eye. She also had a right relative afferent pupillary defect and swelling of her right optic disc. The rest of her neurological and ophthalmological exams were unremarkable. She received 5 days of high dose intravenous methylprednisolone (IVMP), with mild improvement in her

vision. One month later, she had painful deterioration in her right sight, with eye exam showing vision of 20/200 on the right and optic nerve pallor on the same side. She received another 5 day course of IVMP, with no improvement. She had a third episode of pain upon right eye movement 6 months later, with an unchanged neuro-ophthalmologic exam; she received 3 days of IVMP with no improvement, and developed a central scotoma ever since with best corrected vision of 20/200 on the right and 20/20 on the left. Her neurological and rheumatological review of systems were only remarkable for mild low back pain, with no stiffness or decreased range of motion. She denied alcohol use and reported being in a monogamous relationship with her husband.

Her ocular workup included fluorescein angiography that initially showed leakage from the optic nerve but later showed pooling with no signs of vasculitis (Figure 1). Her last ocular computed tomography of the ganglion layer showed thinning in the right eye (Figure 2a) and similarly, the Retinal Nerve Fiber Layer (RNFL) showed atrophy of the right optic nerve with RNFL thickness of 56 micra compared to 97 micra in the left eye (Figure 2b). Her right visual evoked potential (VEP) was absent, and her left VEP showed a P100 latency of 114 sec (normal).

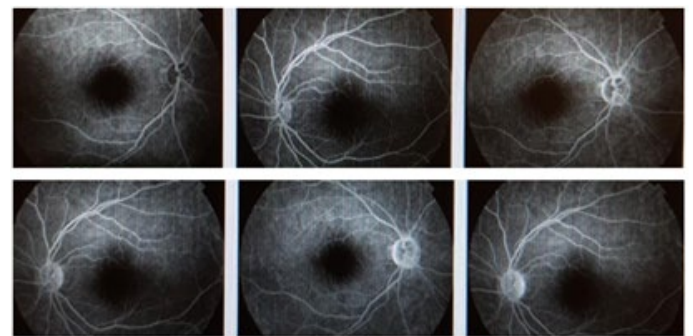


Figure 1. Fluorescein angiography showing no signs of vasculitis.

Brain magnetic resonance imaging (MRI) was unremarkable on the first 2 episodes, but 6 months later showed 2 tiny new gadolinium-enhancing 1 mm cerebellar lesions (Figure 3). Magnetic resonance angiography (MRA) was negative for evidence of vasculitis. Cervical and dorsal MRIs were negative for any intramedullary lesions. Her somatosensory evoked potentials were unremarkable.

Two lumbar punctures were performed, and both cerebrospinal fluid results (CSF) showed no white blood cells, normal protein count and glucose level, negative IgG index and oligoclonal bands. Serologic markers tested included negative aquaporin-4 antibodies, myelin-oligodendrocyte glycoprotein, angiotensin-converting enzyme levels, brucella serology, serum and urine protein electrophoresis and immunofixation, HLA-B52 and antinuclear antibody, anti-double stranded DNA and SSA/SSB. Her HLA-B27 turned out

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to be positive.

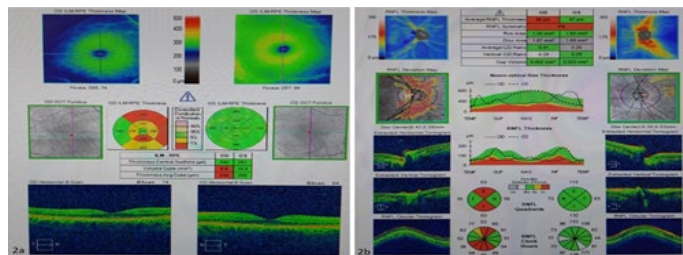


Figure 2. (a) Optical coherence tomography of the ganglion cell layer and (b) The retinal nerve fiber layer.

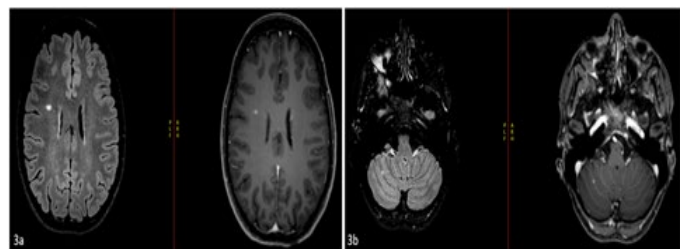


Figure 3. (a) Two high-FLAIR lesions that are enhancing on brain MRI with gadolinium, in the right frontal lobe and (b) The right cerebellum. FLAIR: fluid-attenuated inversion recovery. MRI: magnetic resonance imaging.

Pelvic MRI was negative for sacroiliitis.

Her recurrent isolated right optic neuritis with minimal response to steroids and lack of improvement fits the diagnosis of CRION, associated with HLA-B27 and enhancing brain lesions, one of few described cases in the literature.

She was started on MMF 1 gram twice daily and was clinically and radiologically relapse free since her last episode.

Results and Discussion

Our patient presented with three steroid dependent relapses of optic neuritis, fitting the diagnosis of CRION. Her autoimmune workup was negative, except for HLA-B27 status. ON has been previously associated with HLA-B27. In fact, in a 2016 Chinese retrospective study, 410 patients with ON underwent HLA-B27 screening and 22 patients were found to be positive with mean age of onset 36 years, comparable to our patient's [2]. The majority was women (1.75/1) and the mean disease duration was 36.1 months. Both eyes were involved in 72.7% of patients and 63.6% had recurrent episodes, while only a minority had simultaneous involvement of both eyes. Only 1 patient fit the criteria NMO and 1 for MS. Our patient did not have any clinical or radiological evidence of myelopathy, and had absent aquaporin-4 antibodies in serum, excluding a diagnosis of NMO spectrum disorder. Furthermore, she did not fit the 2017 McDonald's criteria for MS. Her brain MRI showed 2 gadolinium-enhancing lesions in the right frontal lobe and the right cerebellum, highly suggestive of demyelination, which could have been induced by HLA-B27. In fact, myelin basic protein includes specific B27-binding peptides, which have the potential to trigger CNS demyelination through molecular mimicry [3]. Although this patient did not have MS, she needed to be closely monitored for new clinical and imaging findings, as MS has been associated with HLA-B27 and ankylosing spondylitis (AS). AS was ruled out in our patient, with pelvic MRI showing no evidence of sacroiliitis, a highly sensitive finding. CRION, especially in association with HLA-B27, has been shown to affect both eyes, with a relatively high rate of recurrence [2]. Treatment with long-term immunosuppressive agents is crucial to slow down disease progression. MMF

has been proven to control HLA-B27 associated intraocular inflammation, such as uveitis, retinal vasculitis and papillitis, but its efficiency has never been reported in HLA-B27 associated optic neuritis [4]. MMF works by decreasing T and B cell proliferation, as well as their recruitment into sites of inflammation, hence slowing down inflammation related demyelination of the optic nerve and the brain parenchyma [5,6].

Conclusion

In conclusion, we recommend screening CRION patients for HLA-B27, and if positive, follow up with serial brain MRIs to rule out MS is pivotal. Mycophenolate mofetil appears to have a robust anti-inflammatory action in HLA-B27 associated CRION and CNS demyelination, slowing and possibly halting the progression of disabling vision loss and focal neurological deficits.

Ethics Approval

Not applicable.

Consent for Publication

Consent was obtained from the patient.

Data Availability

Not applicable.

Conflict of interest

The authors of this manuscript have no conflicts of interest to report.

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Author's Contributions

The authors equally contributed to this manuscript.

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