Case Report

A Case of Post Encephalitic Optic Neuritis: Clinical Spectrum, Differential Diagnosis and Management

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Abstract

Purpose: Most cases of optic neuritis are idiopathic or are associated with multiple sclerosis. We present a case in which a young female developed post-infectious left optic neuritis following herpes simplex encephalitis (HSE).

Case Report: A 24-year-old female presented with a severe headache, fever, and malaise of a one-week duration. Viral encephalitis was diagnosed and treated; intravenous acyclovir (750 mg every 8 h) was administered for 14 days. The patient improved clinically and was prescribed oral valacyclovir (1,000 mg, three times daily) for an additional 3 months as an outpatient. The patient presented again four weeks after the initial admission with left periocular pain and other typical manifestations of optic neuritis. We diagnosed post-infectious left optic neuritis following viral encephalitis. Corticosteroid therapy with 250 mg intravenous methylprednisolone every 6 hours was initiated and the patient showed rapid significant recovery.

Conclusion: This case report highlights the patient's clinical course and includes a brief history of the systemic effects of HSE, as well as the pathophysiology, management, and differential diagnosis of post-encephalitic optic neuritis. We suggest that clinicians should routinely perform an ophthalmologic examination during the follow-up visits of such patients.

Keywords: Acyclovir; Corticosteroid; Encephalitis; Herpes Simplex Virus; Optic neuritis

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INTRODUCTION

Optic neuritis is defined as painful inflammation of the optic nerve. It is the most common cause of sudden acute unilateral vision loss or reduced vision in children and young adults.[1] In most cases, optic neuritis occurs in association with multiple sclerosis (MS) or as an isolated syndrome due to idiopathic inflammatory demyelination.[2] Although post-infectious optic neuritis is an uncommon clinical manifestation of herpes simplex virus-1 (HSV-1) encephalitis, it can be seen one to three weeks after the symptomatic infective prodromal phase.[3,4] A case of optic neuritis, a rare complication of HSV-1 infection in immunocompetent adults, is reported here; comparisons to published cases in the literature are included.

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CASE REPORT

A 24-year-old female reported the primary complaint of a severe headache and fever of a two-week duration. She presented with a history of a headache in the right and left temporoparietal regions accompanied by fever, malaise, and vomiting, which had become worse over a one-week period. On admission, the patient was found to have an altered mental status without focal weakness. There was no history of photophobia, blurred vision, or diplopia, or any significant past medical history. On physical examination, the patient's vital signs were stable and within normal limits. On clinical examination, Kernig's sign was negative; gait and coordination were normal; and the sensory system, motor system, and cranial nerves were normal. An ophthalmologic examination including fundoscopy did not reveal any abnormality. Other systemic examinations did not reveal any significant findings. Routine complete hematological and biochemical analyses did not reveal any abnormalities. The electroencephalogram (EEG) was normal. However, an enzyme-linked immunosorbent assay (ELISA) of the patient's serum was positive for anti-HSV IgM antibodies. A neurological investigation commenced. Cranial imaging with a computed tomographic (CT) scan was normal; however, magnetic resonance imaging (MRI) of the brain revealed areas of focal edema in the temporal lobes and insular cortex, as well as the angular gyrus. T2-weighted MRI revealed hyperintensity in both temporal regions medially, sparing the basal ganglia [Figure 1]. Axial diffusion-weighted imaging revealed restricted diffusion in the right and left medial temporal lobes consistent with herpes simplex encephalitis (HSE).

Based on the serological and MRI findings, a presumptive diagnosis of HSE was made. HSV-1 disease was confirmed by cerebrospinal fluid (CSF) analysis with polymerase chain reaction (PCR) quantification. Intravenous acyclovir was administered (750 mg every 8 h) for 14 days. The patient improved clinically. A repeat MRI following treatment showed reduced temporal enhancement. Oral valacyclovir (1,000 mg three times daily) was administered for an additional 3 months as an outpatient. The patient presented again four weeks after initial admission with the left periocular pain of a three-day duration and reduced visual acuity in the affected eye. An ophthalmologic examination revealed a swollen and hyperemic left optic disc, impaired color vision, an afferent papillary defect, and progressive vision loss in the affected eye. There was a delay in left P100 latency and reduced visual evoked potential. An MRI of the brain with fat-suppressed postgadolinium T1-weighted images including orbital cuts revealed an intensely enhanced segment in the left optic nerve [Figure 2]. Axial and coronal short tau inversion recovery images [Figures 3a and b] showed a faint increased signal in the left optic nerve. In comparison, the right optic nerve was normal. The MRI findings, history, and clinical spectrum indicated a diagnosis of post-encephalitic left optic neuritis. Prompt corticosteroid therapy of intravenous methylprednisolone (250 mg every 6 hours for 3 days) was initiated, followed by oral prednisolone (1 mg/kg/d) for 11 days and a 3-day tapering dose of prednisolone thereafter. The patient showed significant improvement in color vision, contrast sensitivity, and the visual field, and left eye pain resolved completely.

DISCUSSION

Viral encephalitis is a rare neurological disorder characterized by an acute, usually diffuse, inflammatory process affecting the brain. It can be caused by other

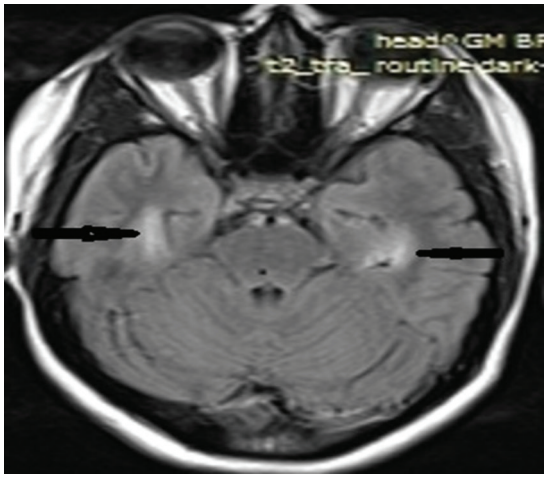

Figure 1. MRI (magnetic resonance imaging) brain FLAIR (fluid attenuation inversion recovery) sequence before treatment showing bilateral hyperintensity in the medial temporal regions (black arrows), suggesting encephalitis.

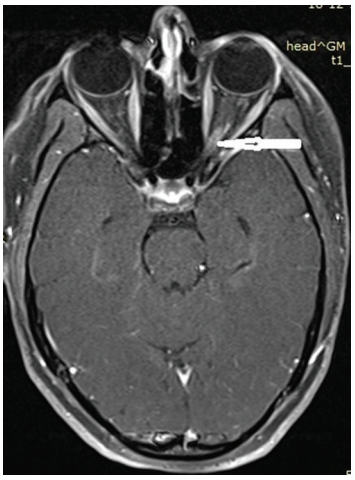

Figure 2. Axial, fat-suppressed, postgadolinium, T1-weighted image including orbital cuts revealed an intensely enhancing segment of the left optic nerve (white arrow).

pathogens, but HSV, a DNA virus, is the most common cause of fatal sporadic encephalitis.[3] HSE is associated with a 70% mortality rate if antiviral therapy is delayed. If HSE is suspected, treatment should be initiated based on a presumptive diagnosis.[5] In the case described here, a definitive diagnosis of HSE was made based on CSF PCR. The resolution of the lesion on MRI following treatment with acyclovir further confirmed the diagnosis. Post-infectious optic neuritis is more common in pediatric patients and usually follows infections such as measles, mumps, and chicken pox, which are more frequent in children.[6] The post-immunization etiology of optic neuritis has also been described.[7,8]

Here, we described a case of viral encephalitis followed by optic neuritis in an immunocompetent adult. The patient presented with the typical acute manifestations of optic neuritis approximately four weeks after the initial hospitalization for encephalitis. Optic nerve involvement after four weeks of initial viral infection suggests a delayed immune response. The delayed onset of optic neuritis after HSE and the complete recovery of visual function, as observed in this case, suggest a reversible process substantiating an immunologic mechanism.

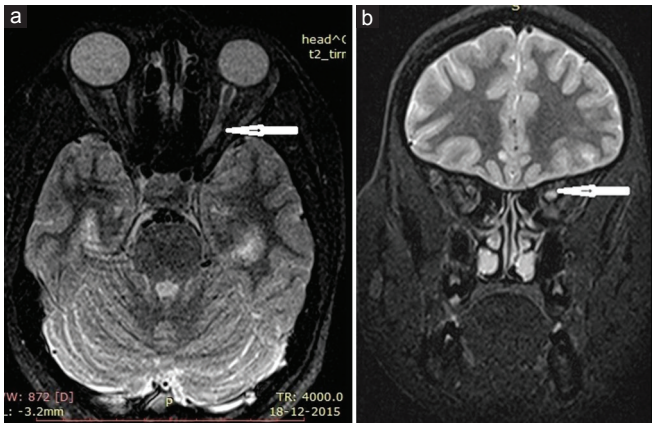

Figure 3. (a) Axial and (b) coronal short tau inversion recovery (STIR) images demonstrated a faint increased signal in the left optic nerve (white arrow). Note the normal right optic nerve for comparison.

The general diagnostic approach to the differential diagnosis consists of a careful history including any recent infection or vaccination, clinical examination, and an investigative profile including general and specifically neurological investigations. Infections that specifically involve the optic nerve, such as *Cryptococcus*, tuberculosis, rickettsia, cerebral malaria, and Lyme disease should be ruled out by routine hematological and biochemical blood screenings. A highly elevated erythrocyte sedimentation rate might suggest tuberculosis or an underlying malignancy, and leucopenia may occur in rickettsial infections. Biochemical screening is helpful in cases of suspected metabolic encephalopathy. Cranial imaging with MRI should be performed, primarily to exclude intracranial abscess, tumors, and demyelinating disorders such as MS or neuromyelitis optica (NMO). Patients should also be evaluated for anti-NMO antibodies, markers for vasculitis, and sarcoidosis. It is also important to distinguish acute disseminated encephalomyelitis as the cause from other conditions, such as acute infectious encephalitis, acute non-infectious encephalitis, and acute metabolic or toxic encephalopathy.[9] The differences are summarized in Table 1 Reproduced from Kennedy,[10] with permission. These features are only guidelines for the clinician to help distinguish between the two differential diagnoses. In the current case, the etiological link between the HSV and optic neuritis can be established indirectly by

Table 1. Comparison of infectious encephalitis and acute disseminated encephalomyelitis (ADEM)

Clinical Features	Infectious Encephalitis	ADEM
Age	Any age	Children
Recent vaccination	Uncommon	Common
Prodromal illness	Occasionally	Usually
Visual loss (one or both eyes)	Uncommon	May occur
Spinal cord signs	Rare	May occur
Routine hematological investigations	Leukocytosis is common	Leucocytosis occasionally occurs
MRI (T2-weighted)	One or more diffuse areas of hyperintensity involve the grey matter of both cerebral cortices, the underlying white matter, and, to a lesser extent, the basal ganglia, brainstem, and cerebellum.	Multiple focal areas of hyperintensity that are the same and may involve the white matter of both hemispheres, the basal ganglia, brainstem, cerebellum, and spinal cord.
CSF analysis	Lymphocytic pleocytosis, elevated protein, normal glucose, and negative cultures. Red blood cells may be seen in herpes simplex encephalitis.	Lymphocytic pleocytosis, elevated protein, normal glucose, and negative cultures. Red blood cells may be seen in acute hemorrhagic leucoencephalitis.

MRI, magnetic resonance imaging; CSF, cerebrospinal fluid.

the history of encephalitis and the exclusion of other common etiologies; a histopathological diagnosis of a direct specimen of the optic nerve is not possible. Optic neuritis is a rare manifestation of HSE, but immediate vigorous treatment with intravenous corticosteroids (in the absence of contraindications and when appropriate) relieves the symptoms, accelerates recovery, and preserves residual visual function.[2,4,6] We suggest that the follow-up visits of such patients should routinely include an ophthalmologic examination including fundoscopy.

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.

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Conflicts of Interest

There are no conflicts of interest.

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