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Demyelinating optic neuritis presenting as a clinically isolated syndrome

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Abstract

BACKGROUND: Clinically isolated syndrome (CIS) describes a single, first-occurrence attack caused by inflammation/demyelination in 1 or more locations in the central nervous system. The optic nerve is a frequent site affected by this neurologic event. As the name implies, CIS is an isolated condition but is often considered a precursor to multiple sclerosis (MS). When distinctive brain lesions detected by magnetic resonance imaging (MRI) accompany CIS, the person is considered at a high risk for MS. Treatment is aimed at delaying the onset of a second neurologic episode, reducing the accumulation of MRI-detected brain lesions and delaying the development of definite MS.

CASE REPORT: This article describes a 40-year-old woman who experienced a sudden loss of vision in the right eye. Testing ultimately found a normal MRI, demyelination of the optic nerve, and progressive thinning of the retinal nerve fiber layer, leading to a diagnosis of CIS.

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Clinically isolated syndrome (CIS) is the first attack on the central nervous system (CNS) indicating inflammation and demyelination. The detection of characteristic brain lesions by neuroimaging suggests a high likelihood for the development of subsequent episodes, often leading to multiple sclerosis (MS).

Case report

A 40-year-old black woman was referred for a neuro-ophthalmologic consultation regarding a sudden vision loss in the right eye that started approximately 3 weeks prior. Since onset, the patient noted pain on eye movement, light sensitivity, and decreased color vision in the right eye. She also reported episodes of intermittent diplopia, dizziness, and frontal headaches. She indicated that her left eye had always been a "lazy eye," and review of her medical records confirmed that she was a refractive amblyope in the left eye. She denied previous episodes of paresthesias, transverse myelitis, or optic neuritis. Her medical history included a diagnosis of Paget's disease (osteitis deformans) 5 years prior, for which she was treated with zoledronic acid (Reclast®; Novartis, Parsippany, New Jersey), calcium, and vitamin D. She had no known allergies, and she denied smoking or alcohol consumption. Her social history and family history were unrevealing.

At the consult visit, the patient's best-corrected visual acuities (BCVA) were 20/40 in each eye. Ocular motilities were full, but the patient noted discomfort in the right eye upon movement. There was no sign of internuclear

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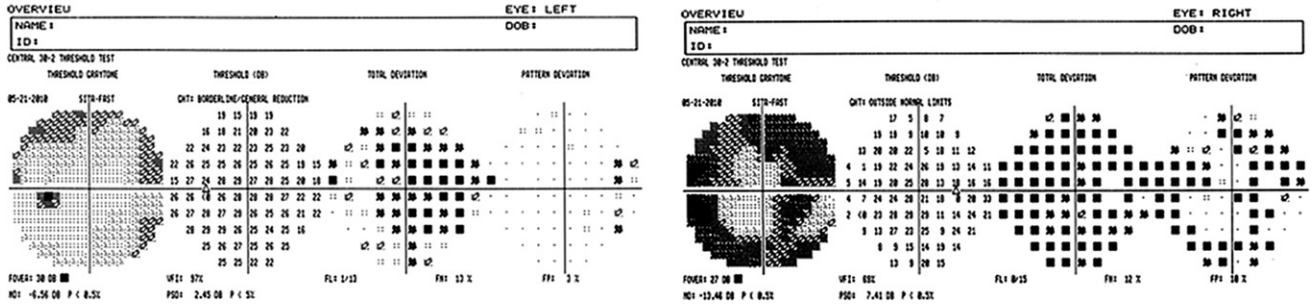

Figure 1 Humphrey visual fields 30-2; this is the baseline test at the initial consult visit.

ophthalmoplegia or nystagmus. Humphrey visual field (30-2) showed peripheral scotomas in both eyes (right eye greater than left eye) but with low reliability (*see* Figure 1). The Amsler grid appeared faded when viewed with the right eye, but there were no distortions reported in either eye. Color vision testing, using Hardy-Rand-Ritter plates, indicated a nonspecific color defect in the right eye, with 13 of 24 plates correctly identified and no color defects in the left eye. A right relative afferent pupillary defect (RAPD) was measured to be 1.2 log units. Intraocular pressures (IOPs), assessed by Goldmann tonometry, were 12 mmHg and 10 mmHg in the right and left eyes, respectively. A fundus examination found flat discs, distinct margins, and temporal pallor of the right optic disc (*see* Figure 2). The macula and other retinal structures were unremarkable in both eyes (*see* Figure 3). There were no retinal nerve fiber layer (RNFL) defects seen on Cirrus optical coherence tomography (OCT; Carl Zeiss Meditec, Dublin, California) (*see* Figure 4), but the macula showed marginal thinning in both eyes (*see* Figure 5). The visual evoked potential (VEP) of the right eye showed a reduced amplitude and delayed implicit time of 45 msec, whereas the left eye was normal (*see* Figure 6). Prior magnetic resonance imaging (MRI) of the brain, obtained by her neurologist shortly after the onset of symptoms, revealed no T2 or contrast-enhancing white matter lesions (*see* Figure 7).

The diagnosis was acute retrobulbar optic neuritis in the right eye. Evidence of demyelination, indicated by the delay on the VEP and normal imaging, suggested CIS. The patient was sent back to her neurologist to consider treatment with intravenous (IV) steroids, and she was scheduled to return in 1 week for further assessment.

By the 1-week progress visit, the patient had received 2 doses of IV steroids (methylprednisolone sodium succinate) and had exhibited rapid recovery. Her BCVAs were now 20/25 in the right eye and 20/30 in the amblyopic left eye. A reliable Humphrey 30-2 showed significant improvement (*see* Figure 8). The right RAPD recovered to 0.3 log units, and the IOPs were consistent with those of the previous visit. The optic neuritis appeared to be responding to treatment, and the diagnosis remained CIS. The patient was continued on a tapering dose of oral prednisone and was scheduled to return in 2 weeks.

At the 3-week follow up, approximately 5 to 6 weeks after onset of symptoms, the BCVA improved to 20/20-1 in the right eye and remained unchanged in the left. Color vision was essentially restored, and Amsler grid testing was normal in both eyes. The right RAPD remained at 0.3 log

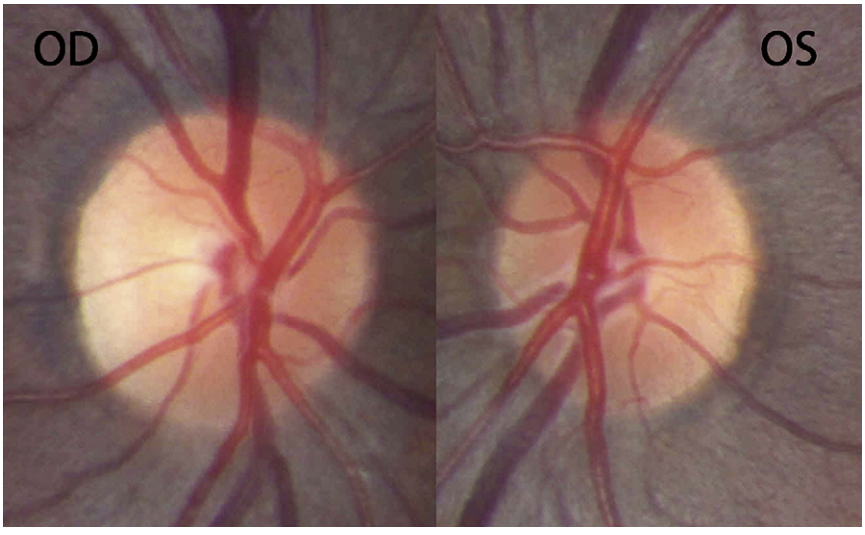

Figure 2 Temporal pallor of the right optic nerve.