ELSEVIER

AFTERIMAGES

JONATHAN WIRTSCHAFTER AND ANDREW HARRISON, EDITORS

Disk Edema and Cranial MRI Optic Nerve Enhancement: How Long is Too Long?

Michael S. Vaphiades, DO

Departments of Ophthalmology and Neurology, University of Arkansas for Medical Sciences, Little Rock, Arkansas, USA

Abstract. A 43-year-old woman presented with painful visual loss and optic disk edema in the right eye (OD) diagnosed as optic neuritis. Initial non–gadolinium-enhanced fat suppressed cranial magnetic resonance imaging (MRI) was normal. Three months later, the disk edema persisted and a gadolinium-enhanced MRI scan of the brain and orbits with fat suppression showed enhancement of the optic nerve OD, most consistent with an optic nerve sheath meningioma. The diagnostic difference between optic neuritis and optic nerve sheath meningioma is discussed. (**Surv Ophthalmol 46:**56–58, 2001.

Key words. disk edema • meningioma • optic nerve sheath meningioma • optic neuritis • orbital MRI with fat suppression and gadolinium

Case Report

A 43-year-old-woman noted discomfort in the right eye (OD) for one week associated with a central scotoma. Her left eye (OS) was normal. She has no medical problems and is taking no medications.

Neuro-ophthalmologic examination on March 3, 1998, showed a visual acuity of 20/20-2 OD and 20/20 OS. Color vision was 5 of 14 OD and 8 of 14 OS, using the Ishihara pseudo-isochromatic plates. Goldmann perimetry revealed an enlarged blindspot OD and normal visual field OS. Pupils were equal with normal reactivity, and there was 0.9 log unit relative afferent pupillary defect OD. Motility findings, trigeminal, and facial function, palpebral fissures, and exophthalmometry were normal. Slit lamp biomicroscopy was normal. Ophthalmoscopy showed optic disk edema OD and a normal-appearing optic nerve OS. The remainder of the retinal examination was normal.

Prior to presentation, a non–gadolinium-enhanced, fat suppressed T1- and T2-weighted magnetic resonance imaging (MRI) scan of the brain and orbits was obtained and was normal (Fig. 1). To better define the lesion, a repeat cranial and orbital MRI with fat suppression and gadolinium was recommended. The patient refused.

Medical evaluation included a complete blood count, electrolytes, antinuclear antibody, syphilis serology, and serum glucose, all of which were normal.

The patient was re-evaluated by her general ophthalmologist on March 30. At that time, she had a visual acuity of 20/200 OD and 20/25 OS and persistent disk edema OD. She was treated with intravenous methylprednisolone 1 g/day for 3 days and a prednisone taper for presumed optic neuritis. She was examined again on April 9, April 13, April 30, May 4, and May 18, 1998. On each visit her visual acuity slowly improved OD; however, the disk edema persisted. A neuro-ophthalmologic examination on

0039-6257/01/$–see front matter
PII S0039-6257(01)00226-0

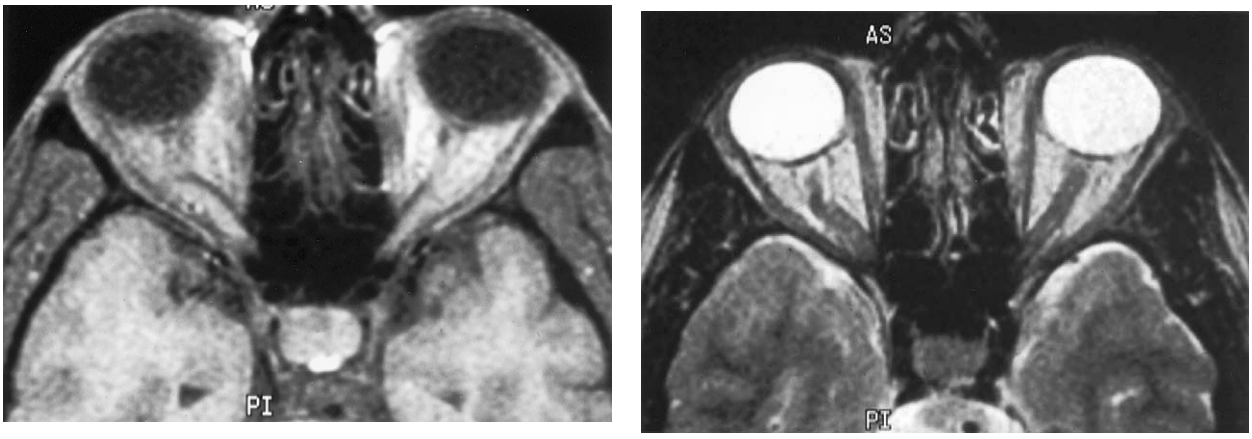

Fig. 1. Initial images. Normal axial fat suppressed non–gadolinium-enhanced orbital T1- and T2-weighted MRI scan.

May 21, 1998, showed visual acuity of 20/30 OD and 20/20 OS. Color vision was 6 of 14 OD and 11 of 14 OS, using the Ishihara pseudo-isochromatic plates. Goldmann perimetry revealed a central scotoma OD and normal visual field OS. Pupils and motility examination were unchanged and ophthalmoscopy continued to show optic disk edema OD with a normal appearing optic nerve OS. The remainder of the retinal examination was normal.

Further testing included a chest radiograph and serum angiotensin converting enzyme (ACE), which were normal. A gadolinium-enhanced T1-weighted MRI of the brain and orbits with fat suppression showed enhancement of the optic nerve OD most consistent with an optic nerve sheath meningioma (ONSM, Fig. 2).

Discussion

Much of our information about optic neuritis is derived from the Optic Neuritis Treatment Trial (ONTT).[1–5,11] In this multicenter study, disk edema occurred in 35.3% of patients at presentation.[2] Much of the visual recovery in optic neuritis occurs after one month, and optic disk pallor occurs in cases with incomplete and complete recovery.[5] In some cases with intermittent follow-up, persistent optic disk edema may really represent recurrent optic neuritis. This was obviously not the case in our patient. Moreover, her optic disk edema did not improve over a period of 3 months, uncharacteristic for optic neuritis.[3] With ONSM, the optic nerve is swollen in 59% of patients, 41% having optic atrophy.[15] Twenty-six percent of patients have optociliary collateral shunt vessels of the optic nerve and 57% of these patients have disk edema.[14]

The majority of patients with optic neuritis are between the ages of 20 and 50 years, and 77% are women.[3] In ONSM, the median age of onset of symptoms is 40 years, and the majority are women.[16] Periorbital pain occurs in 92% of patients with optic neuritis, with pain with eye movement in 87%.[2] Our patient had some periorbital discomfort but not with eye movement. Headaches are occasionally seen in patients with ONSM.[14]

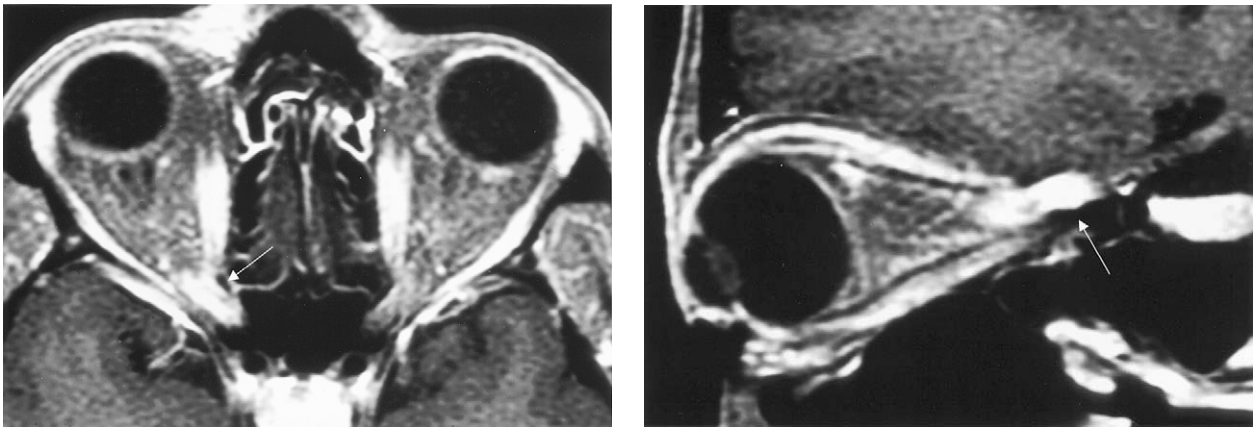

Fig. 2. Afterimages. Axial and sagittal oblique fat suppressed gadolinium-enhanced cranial T1-weighted MRI showing enhancement of the optic nerve OD (arrow) most consistent with an optic nerve sheath meningioma.

Various types of visual field defects may be found in optic neuritis with nerve fiber type defects being most common (20%) and central or cecocentral defects in only 8% of patients.[11] The visual field defects in ONSM include central scotomas, enlarged blindspots, and generalized constriction.[14]

In the ONTT, unenhanced MRI scans were obtained at baseline to evaluate intracranial signal abnormalities (demyelinating plaques). Gadolinium was not generally available and optic nerve imaging was not performed.[4] The best way to evaluate for a demyelinating lesion of the optic nerve is an orbital fat suppressed gadolinium-enhanced cranial MRI.[9] Gadolinium does not cross the blood–brain barrier and enhancement with gadolinium administration denotes disruption of the blood–brain barrier within the optic nerve.[9] The affected optic nerve enhances because gadolinium induces a local magnetic field, resulting in a bright signal on T1-weighted fat suppressed images.[7] Orbital fat suppression is necessary to visualize structures in which there is a large amount of fat, like the orbits.[9,12] This optic nerve enhancement can differentiate between ONSM and optic neuritis. In patients with optic neuritis, gadolinium enhancement is transient, remitting in days. This can differentiate between new and older demyelinating optic nerve lesions.[8] Indeed, optic nerve enhancement for long periods of time can differentiate between ONSM and more remote optic neuritis.[7,8] Cranial and orbital MRI is the best procedure to demonstrate an ONSM.[13] The meningioma appears isointense on T1- and T2-weighted images and enhances diffusely with gadolinium.[13]

A reasonable approach in a patient presenting with a presumed optic neuritis is to obtain a gadolinium-enhanced, orbital fat-suppressed MRI scan of the brain and orbits. This type of MRI is important for two reasons, the first being to rule out other possible causes for the optic neuropathy. The second is to evaluate for lesions consistent with demyelination (including the involved optic nerve), which would support the diagnosis. According to the ONTT, the number of demyelinating lesions on the brain MRI is predictive for the development of clinically definite multiple sclerosis (CDMS).[1] This approach is especially timely, because recent data shows benefit with a weekly treatment of intramuscular interferon beta-1a for patients having their first demyelinating event if they are at high risk for the development of CDMS. This risk is dependent upon the number of brain lesions on the cranial MRI.[10]

If a patient has optic neuritis, follow-up examination within one month should show improvement of the disk edema. At this time, the patient should be re-evaluated for the appearance of a macular star, which may not develop until 2 weeks after presentation of optic disk edema.[6] If a macular star is present, this rules out optic neuritis and the possible development of CDMS, and would prompt a work-up for neuroretinitis.[6]

In summary, if over time the patient's symptoms and examination do not follow the natural history of the presumed diagnosis, one must consider another diagnosis. Cranial MRI with orbital fat suppression and gadolinium enhancement is the procedure of choice when neuroimaging patients with optic neuropathies.

References

1. Optic Neuritis Study Group. The 5-year risk of MS after optic neuritis. Experience of the optic neuritis treatment trial. Neurology 49:1404–13, 1997
2. Optic Neuritis Study Group. The clinical profile of optic neuritis. Experience of the Optic Neuritis Treatment Trial. Arch Ophthalmol 109:1673–8, 1991
3. Beck RW: Optic neuritis, in Miller NR, Newman NJ (eds): Walsh & Hoyt's Clinical Neuro-Ophthalmology, Vol I. Baltimore, Williams & Wilkins, 1998, ed 5, pp 599–647
4. Beck RW, Arrington J, Murtagh FR, et al: Brain magnetic resonance imaging in acute optic neuritis. Experience of the Optic Neuritis Study Group. Arch Neurol 50:841–6, 1993
5. Beck RW, Cleary PA, Backlund JC: The course of visual recovery after optic neuritis. Experience of the Optic Neuritis Treatment Trial. Ophthalmology 101:1771–8, 1994
6. Brazis PW, Lee AG: Optic disk edema with a macular star. Mayo Clin Proc 71:1162–6, 1996
7. Gass A, Barker GJ, MacManus D, et al: High resolution magnetic resonance imaging of the anterior visual pathway in patients with optic neuropathies using fast spin echo and phased array local coils. J Neurol Neurosurg Psychiatry 58:562–9, 1995
8. Guy J, Mancuso A, Quisling RG, et al: Gadolinium-DTPA-enhanced magnetic resonance imaging in optic neuropathies. Ophthalmology 97:592-9; discussion 599–600, 1990
9. Guy J, Mao J, Bidgood WD Jr, et al: Enhancement and demyelination of the intraorbital optic nerve. Fat suppression magnetic resonance imaging. Ophthalmology 99:713–9, 1992
10. Jacobs LD, Beck RW, Simon JH, et al: Intramuscular interferon beta-1a therapy initiated during a first demyelinating event in multiple sclerosis. N Engl J Med 343:898–904, 2000
11. Keltner JL, Johnson CA, Spurr JO, Beck RW: Baseline visual field profile of optic neuritis. The experience of the optic neuritis treatment trial. Optic Neuritis Study Group. Arch Ophthalmol 111:231–4, 1993
12. Lee DH, Simon JH, Szumowski J, et al: Optic neuritis and orbital lesions: lipid-suppressed chemical shift MR imaging. Radiology 179:543–6, 1991
13. Lindblom B, Truwit CL, Hoyt WF: Optic nerve sheath meningioma. Definition of intraorbital, intracanalicular, and intracranial components with magnetic resonance imaging. Ophthalmology 99:560–6, 1992
14. Sarkies NJ: Optic nerve sheath meningioma: diagnostic features and therapeutic alternatives. Eye 1:597–602, 1987
15. Sibony PA, Krauss HR, Kennerdell JS, et al: Optic nerve sheath meningiomas. Clinical manifestations. Ophthalmology 91:1313–26, 1984
16. Wright JE, McNab AA, McDonald WI: Primary optic nerve sheath meningioma. Br J Ophthalmol 73:960–6, 1989

This work was supported in part by an unrestricted grant from the Research to Prevent Blindness, Inc. New York, New York.

The author has no proprietary or commercial interest in any product or concept discussed in this article.

Reprint address: Michael S. Vaphiades, DO, Harvey & Bernice Jones Eye Institute, Department of Ophthalmology, University of Arkansas for Medical Sciences, 4301 West Markham, Mail Slot 523, Little Rock, Arkansas 72205-7199 USA.