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# MR of Vasculitis-Induced Optic Neuropathy

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**PURPOSE:** To describe the MR characteristics of optic neuropathy caused by vasculitis. **METHODS:** Nine cases of optic neuropathy with diagnosis of vasculitis (six with systemic lupus erythematosus and one each with rheumatoid arthritis, Sjögren disease, and radiation vasculitis) were reviewed retrospectively. Patients were 31 to 62 years old, and all but one were women. All patients had MR imaging through the orbits and anterior visual pathways, five with fat suppression, with and without gadopentetate dimeglumine. Five patients also had MR imaging of the entire brain. The size and enhancement of various segments of the optic nerve and anterior visual pathways were studied. **RESULTS:** MR imaging with contrast material showed enhancement and enlargement of segments of the optic nerves and/or chiasm in six of the nine patients (all but three with systemic lupus erythematosus). Enlargement of a segment of the anterior visual pathway never occurred without enhancement, but enhancement alone did occur in three cases. Of the five patients who had MR imaging of the whole brain, abnormalities were seen in three: periventricular hyperintensity in two and a lacunar infarct in one; none had vessel abnormalities. **CONCLUSION:** Because the MR enhancement seen represents disruption of the blood-brain barrier within the optic nerve, MR imaging with gadopentetate dimeglumine and fat suppression should be performed to detect increased permeability of the blood-brain barrier in acute optic neuropathy.

**Index terms:** Vasculitis; Neuropathy; Nerves, optic (II); Eyes, magnetic resonance

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*Optic neuritis* is a term used to describe inflammation, infection, or demyelination of the optic nerve. Optic neuritis classically refers to a clinical disorder characterized by sudden loss of vision in one or both eyes that may or may not be associated with optic disk swelling. Demyelination is by far the most common cause of optic neuritis; however, differentiation from other less common inflammatory conditions cannot be made by ophthalmoscopic appearance alone. The magnetic resonance (MR) imaging abnormalities of the retrobulbar optic nerve and chiasm are seen in 54% to 84% (depending on the series) of cases of demyelinat-

ing optic neuritis but are not specific for its origin (1-4). The purpose of this study was to describe the MR imaging characteristics of optic neuropathy caused by vasculitis, a heterogeneous group of central nervous system (CNS) disorders characterized by inflammation and necrosis of blood vessel walls.

## Materials and Methods

We undertook a retrospective analysis of nine patients with optic neuropathy and a diagnosis of vasculitis (systemic lupus erythematosus [SLE] in six patients, rheumatoid arthritis in one patient, Sjögren disease in one patient, and radiation vasculitis in one patient). These patients had clinical evidence of vasculitis on the basis of serologic testing, history of systemic disease, or previous radiation therapy without evidence of recurrent tumor. MR images were obtained before treatment with corticosteroids in all but two patients (cases 1 and 5).

MR imaging was performed with a 1.5-T MR system with the use of a head coil. MR images through the orbits and anterior visual pathways were obtained in the coronal and axial planes using spin-echo T1-weighted sequences of 550-600/20-33/2 (repetition time/echo time/excitations). Fat-suppression images were obtained with a frequency-selective presaturation pulse in the coronal plane

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TABLE 1: Clinical profile of nine patients with optic neuropathy

Case	Age, y	Sex	Visual Acuity		Optic Disk		Clinical Diagnosis	Laboratory Findings
			R Eye	L Eye	R Eye	L Eye		
1	32	F	CF	CF	Atrophic	Normal	SLE	ANA 1:640 Anti-ds DNA 210 (nl <100) Anticardiolipin increased
2	55	M	NLP	CF	Normal	Normal	SLE	ANA 1:80 Anti-ds DNA 19.3 (nl < 1.0) Anticardiolipin nl
3	59	F	20/20	20/50	Normal	Pallor	SLE	ANA 1:80 Anti-ds DNA 2.2 (nl < 1.0)
4	59	F	20/400	20/30	Normal	Normal	SLE	Anti-ds DNA 29.9 (nl < 1.0) Anticardiolipin nl
5	31	F	20/20	20/15	DE	Normal	SLE	ANA 1:80 Anti-ds DNA 156 (nl < 100)
6	51	F	20/20	CF	Normal	DE	SLE A	ANA 1:1280 Anti-ds DNA 34.1 (nl < 1) Anticardiolipin nl
7	46	F	20/20	20/70, NLP	Normal	Pallor	Postradiation	...
8	54	F	NLP	20/25	Normal	Normal	Rheumatoid arthritis	...
9	62	F	CF	20/30	Pallor	Normal	Sjögren disease	...

Note.—CF = counting fingers, SLE = systemic lupus erythematosus, ANA = antinuclear antibody, Anti-ds DNA = anti-double-stranded DNA antibody, DE = disk edema, NLP = no light perception, nl = normal.

in four patients (cases 1, 3, 8, and 9) and in the axial plane in one patient (case 3). Sagittal T1-weighted images were obtained in only two patients (cases 8 and 9). All these sequences were obtained before and after administration of gadopentate dimeglumine at a concentration of 0.1 mmol/kg body weight. Section thickness was 3 mm with an 18-cm field of view.

MR images of the entire brain were also obtained in five of the nine patients (cases 1, 2, 6, 8, and 9). This included dual-echo axial images as well as T1-weighted images in axial, coronal, and sagittal planes after the administration of gadopentate dimeglumine at a concentration of 0.1 mmol/kg body weight. T1-weighted coronal images of the brain without the administration of contrast material were also obtained.

The optic nerve and anterior visual pathways were divided into the following segments: intraorbital optic nerve, intracanalicular optic nerve, intracranial optic nerve, chiasm, and optic tract. The size and enhancement of the various segments were studied.

## Results

The demographic and clinical data of each patient are summarized in Table 1. The MR findings of each patient are summarized in Table 2.

Three (50%) of six patients with SLE had abnormalities on MR images. One patient (case 1) had enhancement of the intracranial optic nerves and chiasm as well as enlargement of the chiasm (Fig 1). A second patient (case 2) had enhancement of the intracanalicular left optic nerve, enhancement and enlargement of the

intracranial left optic nerve, and enhancement of the left lateral chiasm and left optic tract. In case 3, enhancement and enlargement of the intraorbital left optic nerve was seen (Fig 2). The patient with active radiation optic neuropathy (case 7) had appropriate enhancement and enlargement of the intracanalicular, intracranial optic nerves and chiasm. A single patient (case 8) with rheumatoid arthritis, systemic necrotizing vasculitis, and visual loss had enhancement and enlargement of the intracanalicular and intracranial optic nerves and optic chiasm on the clinically affected side (Fig 3). The patient with Sjögren disease (case 9) (Fig 4) had enhancement and enlargement of the right intracanalicular and right intracranial optic nerve as well as the right chiasm. This patient also had enhancement of the left intracranial optic nerve and left chiasm.

Enlargement of any segment of the anterior visual pathway was never detected without concomitant enhancement of that portion of the pathway but enhancement alone of any segment was not infrequent (cases 1, 2, 9).

MR images of the brain were obtained in five of the nine patients. Two of these patients (cases 2 and 9) had T2 hyperintensities in the periventricular distribution. One patient showed a lacunar infarct in the region of the left caudate (case 6). The other two cases (1 and 8) did not have any abnormal signal abnormalities of the

TABLE 2: Sites of contrast enhancement/enlargement

Case	Intraorbital Optic Nerve				Intracranial Optic Nerve				Intracranial Optic Nerve				Chiasm		Tract	
	R Eye		L Eye		R Eye		L Eye		R Eye		L Eye		Enh	Enl	Enh	Enl
	Enh	Enl	Enh	Enl	Enh	Enl	Enh	Enl	Enh	Enl	Enh	Enl				
1	—	—	—	—	—	—	—	—	+	—	+	—	+B	+B	—	—
2	—	—	—	—	—	—	+	—	—	—	+	+	+L	—	+L	—
3	—	—	+	+	—	—	—	—	—	—	—	—	—	—	—	—
4	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—
5	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—
6	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—	—
7	—	—	—	—	—	—	+	+	—	—	+	+	+L	+L	—	—
8	—	—	—	—	+	+	—	—	+	+	—	—	+R	+R	—	—
9	—	—	—	—	+	+	—	—	+	+	+	—	+B	+R	—	—

Note.—*Enh* indicates enhancement; *Enl*, enlargement; *B*, bilateral; +, present; and —, absent.

brain parenchyma. None of the patients had abnormalities of the vessels on MR images of the brain.

## Discussion

Optic neuropathy is any visual loss of optic nerve origin regardless of causative factors. Optic neuritis more specifically is a clinical disorder with visual loss of an acute onset (4 to 7 days) usually associated with pain on eye movement with signs of optic nerve or chiasmal field loss and pupillary changes. Numerous origins may be associated but multiple sclerosis remains the most frequent. Vasculitis as a cause

of optic neuritis is often discussed but remains controversial in its significance (5).

In the past, the use of neuroimaging procedures such as computed tomography in a patient with the diagnosis of optic neuritis and optic neuropathy was to exclude compressive lesions of the optic nerve and optic chiasm. MR imaging has the additional advantage of being able to image intrinsic lesions of the optic nerve, optic chiasm, and tract, as well as lesions involving other white matter tracts.

MR imaging with gadopentetate dimeglumine showed enhancement of the optic nerves and/or chiasm in six of nine patients in our study who had vasculitis-induced optic neurop-

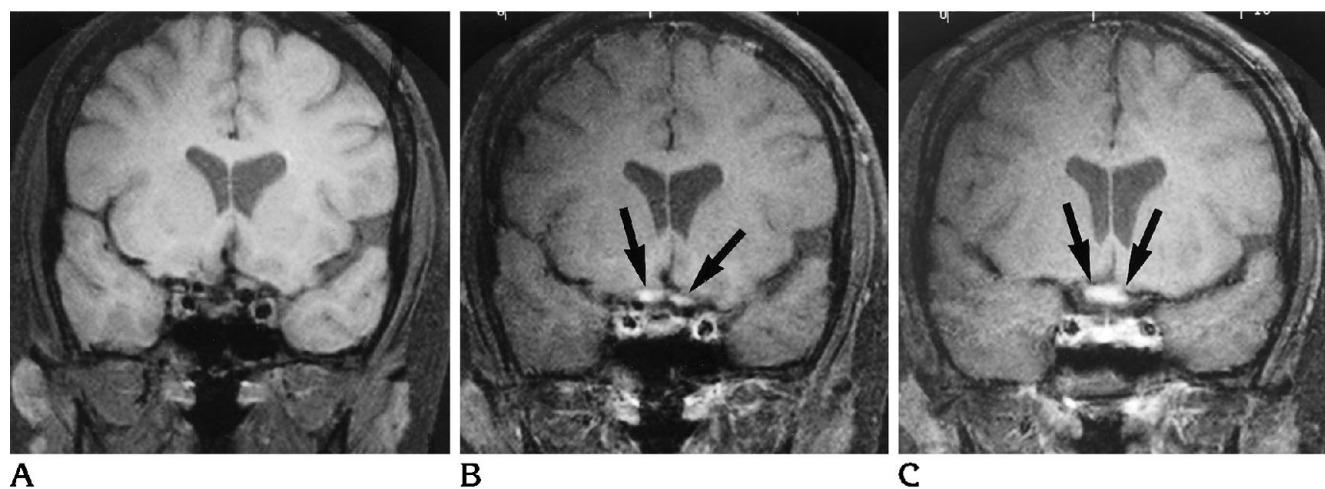


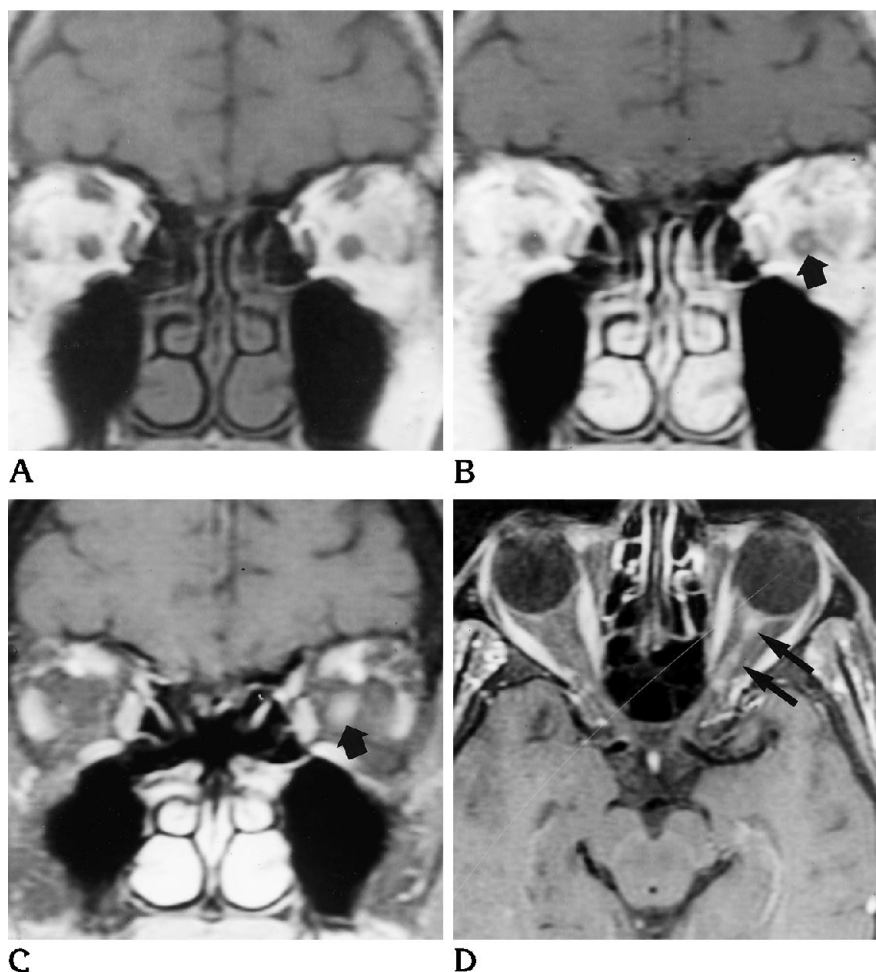
Fig 1. 32-year-old woman (case 1) with a clinical diagnosis of SLE who had prior episodes of visual loss in both eyes. After treatment with corticosteroids, she returned 4 months later with worsening of vision to counting fingers in both eyes, and optic disks were atrophic. A and B, Coronal T1-weighted images before (A) and after (B) administration of gadopentetate dimeglumine show enhancement of the intracranial optic nerves (arrows in B), which were not enlarged.

C, Coronal postcontrast T1-weighted image slightly posterior to those in A and B shows an enhancing and mildly enlarged optic chiasm (arrows).

Fig 2. 59-year-old woman (case 3) with a clinical diagnosis of SLE in whom painful decreasing vision of left eye developed.

A and B, Coronal T1-weighted images before (A) and after (B) administration of gadopentetate dimeglumine show subtle enhancement of the left intraorbital optic nerve (arrow in B).

C and D, In contrast, coronal (C) and axial (D) fat-suppression images after administration of gadopentetate dimeglumine show enhancing structures to better advantage. Enhancing extraocular muscles and left intraorbital optic nerve (arrows) are well seen, unlike the unenhancing right intraorbital optic nerve (courtesy of Thomas P. Naidich, MD, Baptist Hospital, Miami, Fla).



athy. This finding represents disruption of the blood-brain barrier within the optic nerve.

Neurologic involvement in SLE has been described extensively in the literature (5–8) and is responsible for much morbidity and mortality in patients with SLE. Stroke occurs in up to half of the patients and most likely results from cardiac valvular disease, coagulopathy, or antiphospholipid antibody syndrome. Immune complex-mediated cerebral vasculitis is associated with SLE but is uncommon. Angiographic findings range from disease of the normal and small vessels to arteriopathy of the large vessels (9). In their review of the neuropathologic findings in 57 cases of SLE, Ellis and Verity (10) found evidence of cerebral vasculitis in only 7%. Immunoreactants in cerebral vessels and/or the choroid plexus may compromise the integrity of tight junctions of endothelial cells of small vessels in the brain and the epithelial cells associated with the choroid plexus (11). This may alter the permeability of the blood-brain barrier.

An alternative hypothesis in the pathogenesis of neurologic complications in SLE is the involvement of antineuronal antibodies (11). The presence of these antibodies was first suggested by the observation of antilymphocyte antibodies that cross-reacted with neuronal cells (12).

A significant association exists between CNS involvement in lupus and the presence of thrombogenic antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies) (5, 13, 14). These autoantibodies are associated with both CNS and ocular vasoocclusive phenomena (5).

MR findings in SLE patients with nonfocal symptoms include multifocal areas of increased signal intensity in the subcortical white matter on T2-weighted images. Patients with symptoms of focal intracranial disease may have areas of increased signal intensity on T2-weighted images and atrophic changes in regions corresponding to major vascular distributions (15). Multifocal subcortical hemorrhages have been

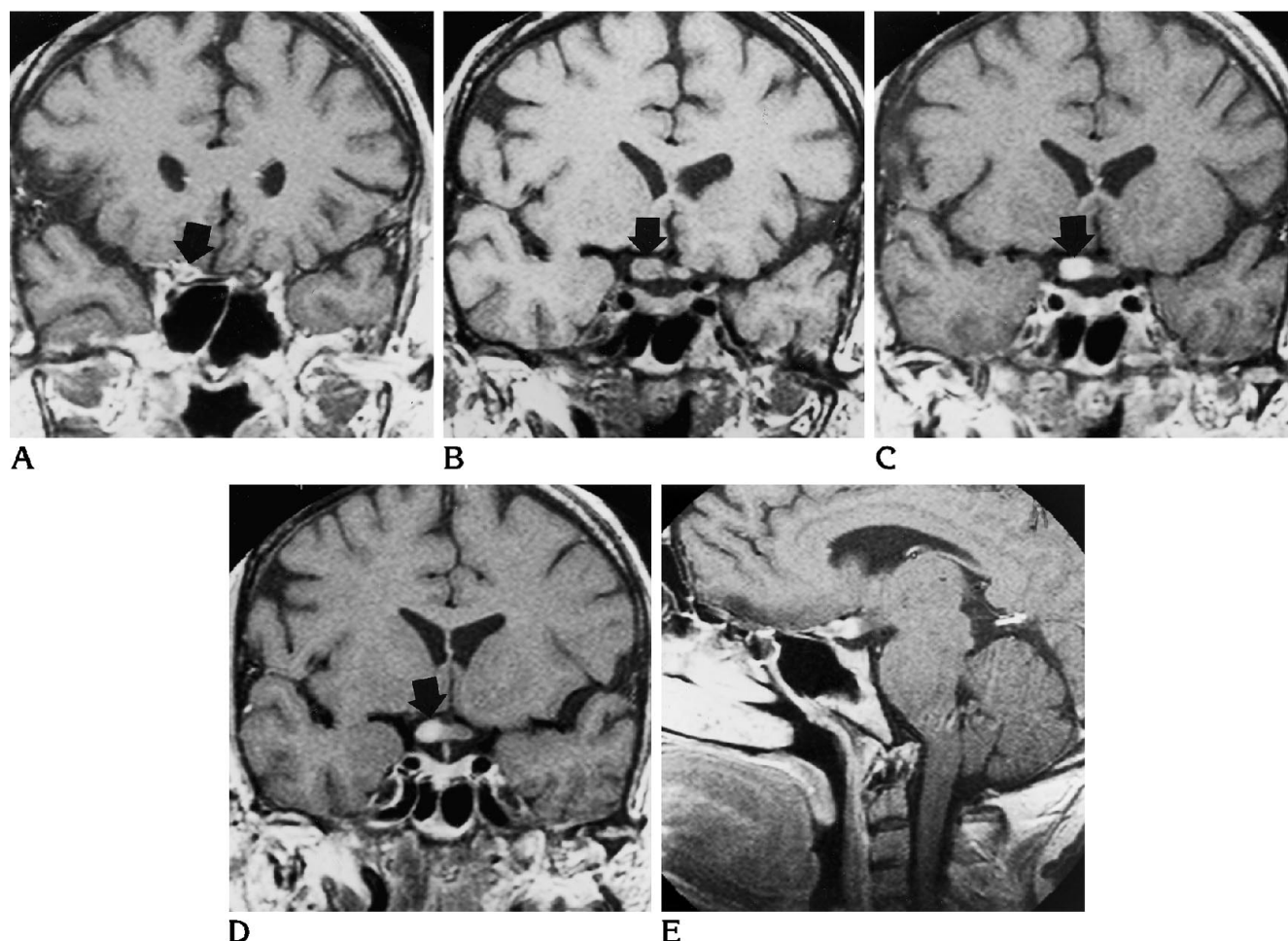


Fig 3. 54-year-old woman (case 8) with severe rheumatoid arthritis who presented with a 5-day history of loss of vision in the right eye.

A, Postcontrast T1-weighted coronal image at the level of the optic canal shows enhancement and enlargement of the right optic nerve just posterior to the canal (*arrow*).

B and C, Slightly more posterior coronal T1-weighted sections before (B) and after (C) administration of gadopentetate dimeglumine show enhancement and enlargement of the right intracranial optic nerve (*arrows*).

D, Postcontrast T1-weighted coronal image through the chiasm shows enhancement and enlargement of the optic chiasm in its right lateral aspect (*arrow*).

E, Postcontrast T1-weighted sagittal image through the right optic nerve shows enhancement and enlargement of the right prechiasmatic optic nerve.

noted as have intracranial calcifications in the basal ganglia and dentate nuclei (16, 17).

SLE can affect many ocular structures. Ocular complications tend to occur in acutely ill patients with active systemic disease (5), and the retina is the most often involved site in the eye (5). Retinal vascular occlusive disease is a devastating complication of SLE (5). The precise pathophysiology underlying the vasculopathy of lupus in the eye is uncertain, but autoimmune mechanisms may play a critical role. Histopathologic studies have shown occlusion of retinal and choroidal vessels (5). Although a true vasculitis is occasionally noted, most oc-

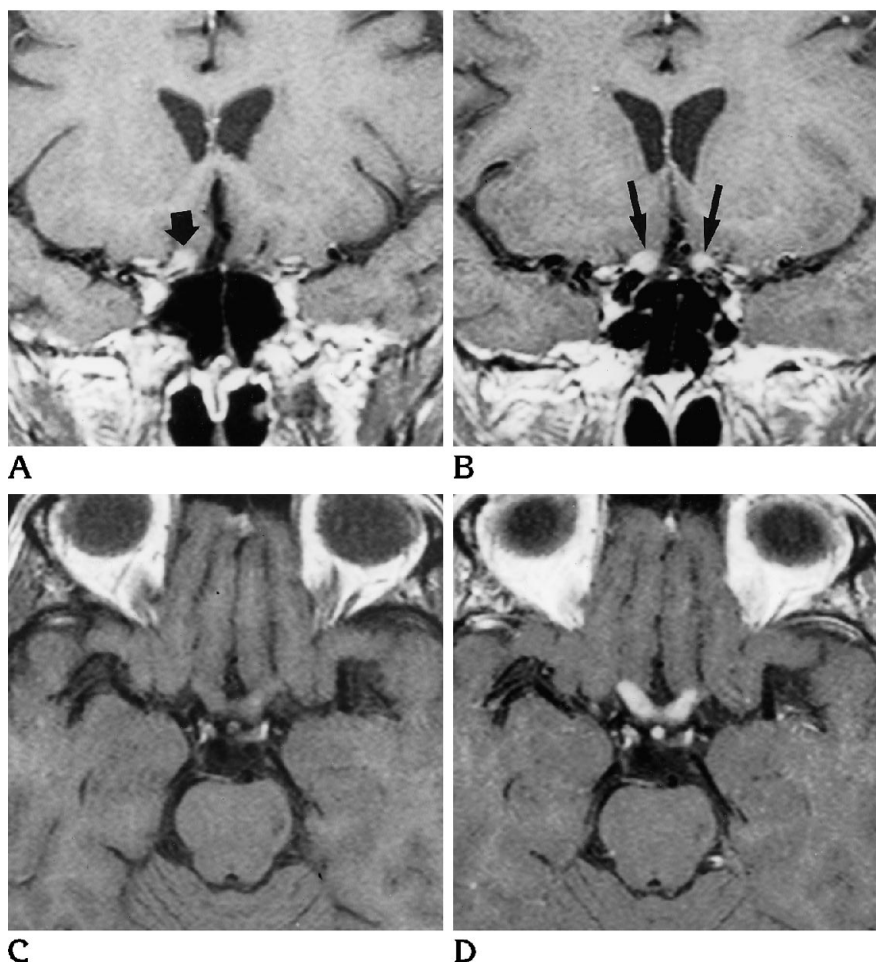
cluded vessels have only a mild perivascular inflammation. Focal fibrinoid necrosis of arterioles with perivascular lymphocyte and plasma cell infiltration may be seen (8). Immune reactants are seen deposited in blood vessel walls within the retina, choroid, ciliary body, sclera, conjunctiva, and corneal epithelium (5). Vessels containing immune deposits show a variety of changes, including thrombosis, endothelial swelling, and focal perivasculitis. Direct involvement of the optic nerve in SLE can occur as acute retrobulbar neuritis, acute anterior optic neuritis, anterior ischemic optic neuropathy, or slowly progressive visual loss (5). The optic

Fig 4. 62-year-old woman (case 9) with a history of Sjögren disease presented with progressive visual loss of the right eye.

A, Postcontrast T1-weighted coronal image shows enhancement and mild enlargement of the right optic nerve just posterior to the canal (*arrow*).

B, Slightly more posterior postcontrast T1-weighted coronal image shows bilateral enhancement of the intracranial portion of the optic nerves (*arrows*) and mild enlargement of the right intracranial optic nerve.

C and D, T1-weighted axial images before (C) and after (D) administration of gadopentetate dimeglumine again show the enhancement of the bilateral intracranial optic nerves and chiasm and mild enlargement of the right intracranial optic nerve.



nerve involvement may be occult or may be the presenting sign of disease.

To make the diagnosis of SLE clinically, four or more of 11 (American Rheumatoid Association) criteria need to be present serially or simultaneously (18). In our study three patients had signs and symptoms that complied with the strict definition of SLE, but SLE was the rational diagnosis in the other three patients who did not fulfill the criteria but who had positive antinuclear antibody titer and anti-double-stranded DNA antibodies.

Involvement of the visual pathways consequent to radiation therapy vasculitis is rare but may occur if total doses exceeding 4500 to 6500 Gy are administered (19–21). Visual symptoms generally occur between 4 months and 3 years after completion of therapy, with the majority of cases occurring 1 year after treatment. Pathologically, reactive gliosis, demyelination, fibrinoid necrosis, and endothelial hyperplasia leading to infarction are seen (22).

In our case of radiation-induced optic neuropathy, recurrent tumor was excluded as a possibility, since the patient's primary tumor (nasal melanoma) was distant from the visual pathway and the time course was good for radiation-induced changes.

The enhancement of the optic nerves and chiasm seen in cases 8 and 9 again represents disruption of the blood-brain barrier. We postulate that this disruption of the blood-brain barrier is vasculitic and may be histopathologically similar to SLE and radiation-induced vasculitis. The patient with rheumatoid arthritis had severe systemic vasculitis with ischemic necrosis of her toes and the patient with Sjögren disease had extensive ischemic demyelination of the deep white matter of the brain.

Suppression of the signal from the orbital fat by techniques such as chemical-shift imaging has been shown to increase detection of lesions involving the intraorbital optic nerve (1–3). In our series only one (case 3, Fig 2) of nine pa-

tients had enhancement of the intraorbital optic nerve. Fat suppression, however, was only used in four of our patients (cases 1, 3, 8, 9). One of these patients had intraorbital optic nerve enhancement. Use of fat-suppression techniques might increase the detection of lesions involving the intraorbital optic nerve in all these patients.

In most cases of optic neuritis, findings on conventional spin-echo MR images are normal, but occasionally focal high-signal-intensity lesions are seen on long-repetition-time images (23). Some investigators have noted a higher sensitivity for detecting optic neuritis on MR images by using inversion-recovery sequences with short inversion times (24). Other authors have recommended MR imaging with gadopentetate dimeglumine as the neuroradiologic procedure of choice for visualization of increased permeability of the blood-brain barrier in acute optic neuritis (4). More recently, fat-suppression techniques in combination with gadopentetate dimeglumine on T1-weighted sequences have been found to improve the delineation of enhancing optic nerve lesions and are superior to contrast-enhanced T1-weighted sequences without fat suppression (1–3). This technique increases the detection of lesions involving the intraorbital optic nerve but has a slight increase in susceptibility effects at the sinus, air, and brain interfaces (1) and therefore may as well be limited to detection of intracranial optic nerve and optic chiasm lesions. Imaging with a surface coil gives excellent delineation of the intraorbital optic nerve, especially when using coronal sections with fat-suppression technique and intravenous contrast material (25). The intracanalicular and intracranial segments of the optic nerve are best seen with the use of a head coil and intravenous contrast material in both axial and coronal sections.

MR imaging with gadopentetate dimeglumine may detect early changes in permeability of the blood-brain barrier before the appearance of neuroophthalmic and pathologic events (5). The combination of history, enhancement, and enlargement of the optic nerve, chiasm, and optic tract may be a clue for further serologic studies. Therapy with corticosteroid or other immunosuppressive agents administered at the appearance of recurrent or new lesions demonstrable on MR images may restore integrity of the blood-brain barrier before irreversible visual loss occurs.

Vasculitis is a rare but important cause of optic neuropathy that recently has been recognized with more frequency. It should always be a consideration in the differential diagnosis of patients with enhancement of the optic nerve and chiasm. MR imaging with gadopentetate dimeglumine as well as with fat-suppression techniques should be the procedure of choice for detection of increased permeability of the blood-brain barrier in acute optic neuropathy.

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