Optic Nerve Sarcoidosis: MR Findings

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Summary: Neurologic involvement has been described in 5% of patients with sarcoidosis; however, direct granulomatous involvement limited to the optic nerve but not involving the chiasm is rare. We describe two patients with biopsy-proven sarcoidosis; the fat-suppressed gadolinium-enhanced MR findings showed sarcoid involvement limited to the optic nerves.

Index terms: Sarcoidosis; Nerves, optic (II)

Neurologic involvement has been described in 5% of patients with sarcoidosis (1); however, direct granulomatous involvement limited to the optic nerve but not involving the chiasm is rare (2). We describe two patients with biopsy-proven sarcoidosis; the fat-suppressed (3) gadolinium-enhanced magnetic resonance (MR) findings showed sarcoid involvement limited to the optic nerves.

Case Reports

Case 1

A 52-year-old white woman noted decreased vision in her right eye and developed complete blindness in her right eye during a 3-week period. Her vision failed to improve during the next 14 months. Short tau inversion recovery (STIR) images showed no significant abnormalities. MR images using fat-suppression techniques revealed enhancement of the right optic nerve without involvement of the left optic nerve or optic chiasm (Fig. 1). She showed no other evidence of systemic sarcoidosis; however, a biopsy of the right optic nerve proper showed noncaseating granulomas confirming the diagnosis of sarcoidosis. She eventually lost all vision in her right eye due to neovascular glaucoma from an occlusion of the central retinal vein.

Case 2

A 43-year-old black woman presented with a 1-month history of right frontal headaches and progressive bilateral visual loss. Her conjunctiva were nodular bilaterally, but the remainder of her ophthalmologic examination was essentially within normal limits. A head computed tomography (CT) scan revealed no abnormal findings. MR images (STIR) showed the optic nerve/sheath complex to be enlarged bilaterally; however, it was not possible to distinguish between the cerebrospinal fluid in the subarachnoid space and the optic nerve proper. A contrast-enhanced fat-suppressed MR examination showed abnormal optic nerve enlargement and enhancement bilaterally, extending beyond the optic apex intracranially but not involving the chiasm (Fig. 2). No meningeal enhancement was identified. A conjunctival biopsy showed noncaseating granulomas, suggesting involvement with sarcoidosis. She showed no other evidence of systemic sarcoidosis. Prednisone was started, and her vision gradually improved and stabilized over the next 5 weeks. A follow-up fat-suppressed MR examination with gadolinium showed the optic nerves still slightly enlarged and abnormally enhanced despite clinical improvement (images not shown).

Discussion

Sarcoidosis is a multisystem, noncaseating, granulomatous disease, most frequently involving the lung. The signs and symptoms usually improve with steroid therapy (4). Brain parenchymal disease is uncommon (4), with central nervous system involvement occurring in about 5% of all patients. Neurosarcoidosis commonly affects the leptomeninges at the base of the skull. Granulomatous infiltration of the basal meninges frequently leads to compression and invasion of the cranial nerves, especially of the optic chiasm, with resultant cranial nerve palsies. About 1%–5% of all patients with sarcoidosis have disease affecting the optic nerve in some manner. Ocular symptoms may be the only clinically detectable manifestations of sarcoidosis, as seen in case 2, and have been reported to be the presenting symptom in about 20% of patients (5). The chiasm is involved far more frequently than the intraorbital optic nerve, and usually there is con-
Fig. 1. A, Coronal T1-weighted (450/11), fat-suppressed postcontrast image shows abnormal enhancement of the enlarged right optic nerve (large black arrow). The signal intensity of the normal left optic nerve (large white arrow) is similar to that of normal brain parenchyma. Note the low signal intensity of cerebrospinal fluid surrounding the left optic nerve and the normal mildly enhanced ring of meninges (small white arrows). Note the absence of cerebrospinal fluid on the right. The normal rectus muscles show normally intense contrast enhancement.

B, Axial T1-weighted (650/11), fat-suppressed postcontrast image shows irregular enhancement of the right optic nerve near the apex involving the intraconal and intracanicular portions of the optic nerve (open arrows). Again noted are the noninvolved left optic nerve and chiasm (large arrows) showing a similar degree of signal intensity to that of normal brain parenchyma.

Fig. 2. A, Coronal T1-weighted (650/11), fat-suppressed postcontrast image shows significant enhancement of the intraconal portions of both optic nerves. Enlargement of the optic nerves is evidenced by a decreased amount of cerebrospinal fluid in the left subarachnoid space and the absence of cerebrospinal fluid in the right subarachnoid space. Also noted is the mildly enhanced meninges (arrow) seen as a semicircular structure just inferior to the left optic nerve.

B, Axial T1-weighted (650/11), fat-suppressed postcontrast image shows the abnormal enhancement of the enlarged optic nerves bilaterally. The involvement on the right side extends beyond the optic apex into the intracranial portion but does not include the chiasm (images not shown). No intracranial dural meningeal enhancement is noted.

C, Axial STIR (2350/90) image shows absence of fat signal in the intraconal space. Although the optic nerve/sheath complex appears abnormal, the separation between the cerebrospinal fluid and the abnormal optic nerve cannot be identified clearly.
comitant hypothalamic or widespread central nervous system involvement (6). Direct sarcoid granulomatous involvement of the optic nerve is a rare phenomenon (2, 6). In our cases, meningeal or chiasmatic involvement was not evident on MR studies, suggesting direct isolated optic nerve involvement rather than a meningeal process.

CT is not sensitive in the detection of optic nerve lesions near the orbital apex because of beam-hardening artifacts. Generally, enhancement of the optic nerve is not distinct on the spin-echo sequences because of the fatty tissue in the intraconal space, which has a high signal intensity on T1-weighted contrast-enhanced spin-echo images. The abnormal enhancement of the optic nerve can be very difficult to appreciate. Fat-suppression techniques have been reported to be useful in the evaluation of optic nerve pathology with significant improvement in the delineation of optic nerve lesions (7, 8). This technique allows selective reduction of the fat signal intensity without affecting the signal of the adjacent contrast-enhanced tissues. As shown in our cases, the enhancing optic nerve lesions showed significant signal contrast to the surrounding fatty tissue on the coronal fat suppression images (Figs. 1A and 2A) and, therefore, were detected easily. This is not true in the coronal T1- or T2-weighted spin-echo images or STIR images, where the abnormal optic nerve may not be separated readily from the surrounding fatty tissue and/or subarachnoid space. Because of the partial volume effect, axial images may not be the optimal plane for the separation of the optic nerve from surrounding structures.

In conclusion, fat-suppressed contrast-enhanced MR examinations appear to be sensitive in the detection of sarcoidosis of the optic nerve, which, although rare, should be included in the differential diagnosis of optic nerve disease, even in patients without other systemic manifestations of the disease. Follow-up MR examinations may demonstrate improvement; however, slight enlargement and enhancement of the optic nerves may persist despite clinical improvement, as in case 2.

References