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F O Walker, W T McLean, Jr, A Elster and C Stanton

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Chiasmal Sarcoidosis

Francis O. Walker,[1] William T. McLean, Jr.,[1] Allen Elster,[2] and Constance Stanton[3]

The CNS manifestations of sarcoidosis are protean and present a difficult diagnostic challenge when systemic evidence of the disease is absent [1]. However, the combination of diabetes insipidus and chiasmal dysfunction may be particularly suggestive of neurosarcoidosis [2]. We describe a case of chiasmal sarcoidosis in which there was remarkably specific clinical and pathologic correlation with the findings on contrast-enhanced MR images.

Case Report

A 59-year-old woman developed progressive thirst, polydipsia, and polyuria over the course of 1 month. She had no polyphagia or previous psychiatric history, and her family physician excluded the presence of diabetes mellitus. Her symptoms persisted unabated for 2 months, at which time she developed positive scotomata. She returned to her physician, who diagnosed diabetes insipidus and referred her for further evaluation.

Physical examination revealed an articulate, healthy-appearing woman. She was free of adenopathy, and an eye examination was normal except for the optic disk. Neurologic examination was remarkable for physiologic anisocoria, mild papilledema, corrected visual acuities of 20/50 bilaterally, and no afferent pupillary defect. Strength, eye and face movements, muscle stretch reflexes, smell, sensation, and coordination were normal. Laboratory studies—including chest X-ray, CBC, SMAC, electrolytes, liver function tests, angiotensin converting enzyme levels, antinuclear antibodies, erythrocyte sedimentation rate, mammograms, stool guaiacs, and urinalysis—were all normal. Pulmonary function tests were unremarkable. Serial lumbar punctures revealed normal protein and glucose. CSF mononuclear leukocyte cell counts ranged from 0–10 cell/μl. CSF cytology; cultures for bacteria, fungi, and mycobacteria; and serologic tests for syphilis, cryptococcal antigen, and stains were unremarkable. Tuberculosis skin testing was negative, but controls were normal. Dermatologic examination revealed a small area of cirrhosis; however, the results of a biopsy showed no evidence of inflammation.

Unenhanced T1-weighted MR images showed thickening of the optic chiasm and translocation of the pituitary bright spot to the upper infundibulum (Fig. 1A). Postcontrast (gadopentetate dimeglumine) T1-weighted images revealed robust enhancement of the perichiasmal meninges (Figs. 1B and 1C).

Surgical exposure of the optic nerves for biopsy revealed thickening of the chiasmal arachnoid. This tissue was loose, nonadherent, yet strung out when teased apart with forceps. Neuropathologic examination revealed noncaseating granulomas (Fig. 1D). Stains and cultures showed no evidence of neoplasm or infectious agent.

Postoperatively, after institution of prednisone at 80 mg daily, the patient experienced mild, subjective improvement of vision. She did not develop systemic evidence of sarcoidosis over the ensuing 6 months. Her thirst and water intake diminished dramatically on chlorpropamide (250 mg daily).

Discussion

Sarcoidosis is a systemic inflammatory disorder that can affect the brain parenchyma or meninges [1]. Parenchymal lesions of sarcoidosis are rare and have a nonspecific CT and MR appearance, typically involving multifocal, periventricular, white matter lesions. These do not enhance on CT and have increased signal on T2-weighted MR images; with either technique, they resemble the lesions of multiple sclerosis [3, 4]. Leptomeningeal sarcoid may show contrast enhancement both on CT and MR, and may be indistinguishable from the lesions of meningeal carcinomatosis [5, 6].

In this case, the diagnosis of sarcoidosis showed remarkable clinical and radiologic correlation. Sarcoidosis has a well-recognized predilection for the optic chiasm and hypothalamus [2] that, in this patient, resulted in findings of diabetes insipidus and visual impairment.

MR imaging with contrast enhancement localized the lesion to the chiasmal region, specifically implicating an inflammatory leptomeningeal disease. Biopsy results confirmed the clinical suspicion of sarcoidosis. Of additional interest was the displacement of the posterior pituitary bright spot seen on the precontrast T1-weighted images. This bright spot is associated with the terminal distribution of hypothalamic neuroendocrine outflow into the posterior pituitary [7, 8]. When hypothalamic outflow is obstructed, the bright spot is characteristically displaced in a cephalad direction, a finding that correlates with the presence of diabetes insipidus [9, 10].

This case demonstrates the value of contrast-enhanced MR imaging in chiasmal sarcoidosis. Because isolated CNS sarcoidosis is rare, further studies will be needed to determine

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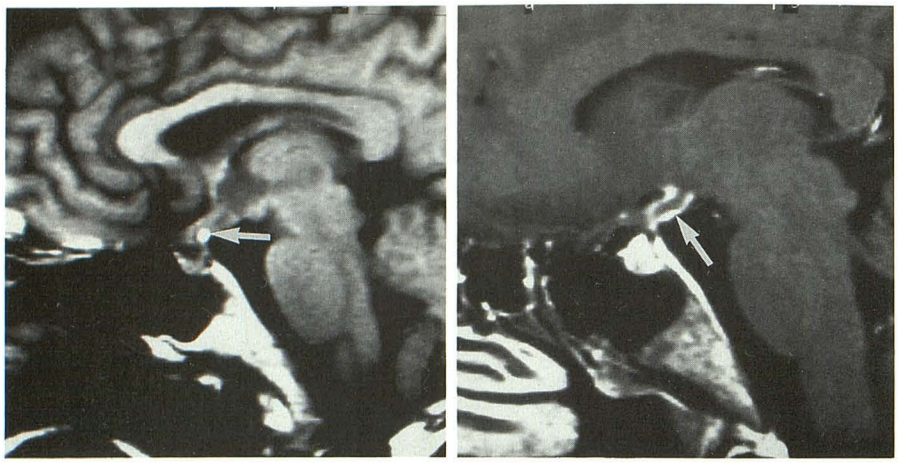
[1] Department of Neurology, Bowman Gray School of Medicine, 300 S. Hawthorne Rd., Winston-Salem, NC 27103. Address reprint requests to F. O. Walker.
[2] Department of Neuroradiology, Bowman Gray School of Medicine, Winston-Salem, NC 27103.
[3] Department of Neuropathology, Bowman Gray School of Medicine, Winston-Salem, NC 27103.



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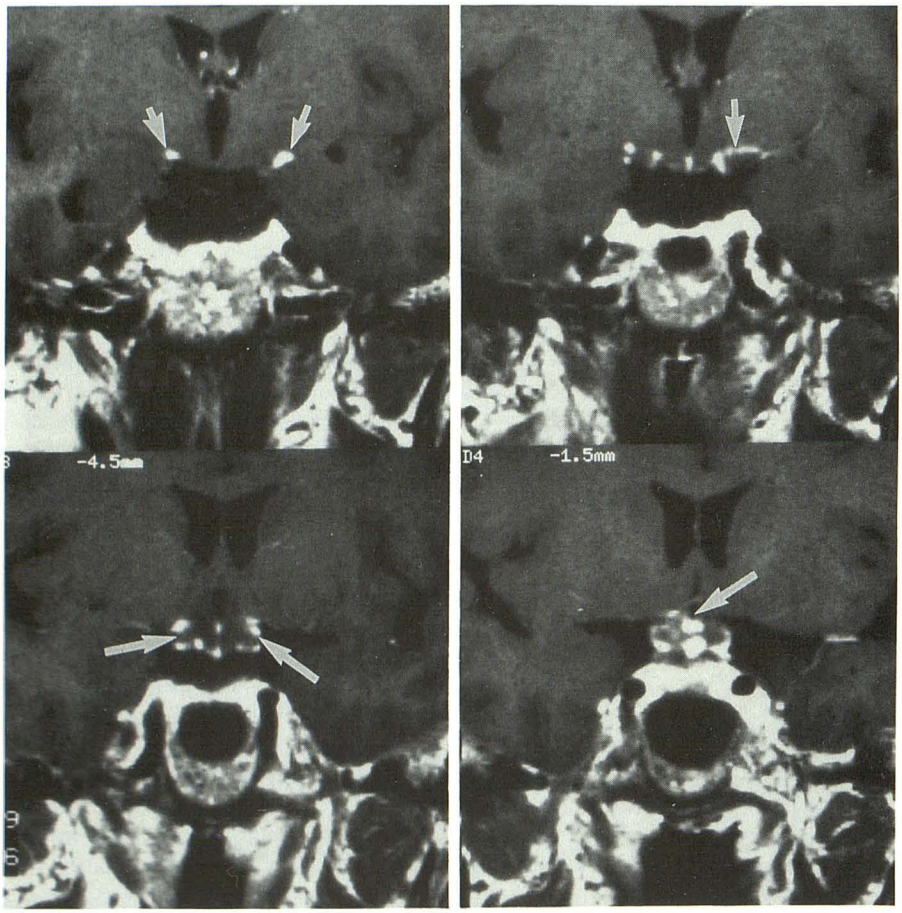

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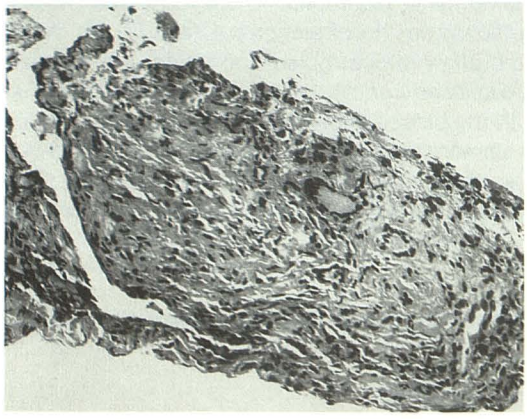

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Fig. 1.—*A*, Precontrast sagittal T1-weighted (500/20) spin-echo MR image shows a normal-size pituitary gland, but rather thick optic chiasm. The pituitary bright spot, normally seen in the posterior lobe, has been translocated to the upper infundibulum (*arrow*).

B, Postcontrast sagittal T1-weighted image shows intense abnormal enhancement of the meningeal lining of the optic chiasm (*arrow*).

C, Multiple postcontrast coronal T1-weighted images show meningeal enhancement around the chiasm, optic tracts, and hypothalamus (*arrows*).

D, Noncaseating granulomas seen on biopsy specimen of the meningeal covering of the optic nerve in this patient. (H and E ×325)

the specificity of this patient's radiologic findings. However, when these findings develop in a patient with known systemic sarcoidosis, they should be considered suggestive of CNS involvement and may constitute sufficient evidence for empiric steroid therapy.

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