Clinical images: prominent perivascular enhancement in primary central nervous system vasculitis

C. Salvarani[3], R.D. Brown Jr[1], J. Huston III[2], G.G. Hunder[4]

From the [1]Department of Neurology and the [2]Department of Radiology, Mayo Clinic, Rochester, Minnesota; [3]Visiting clinician at the Department of Neurology, Mayo Clinic, Rochester, Minnesota, from Unitá Operativa di Reumatologia, Arcispedale S. Maria Nuova, Reggio Emilia, Italy; [4]Emeritus Member, Division of Rheumatology, Mayo Clinic, Rochester, Minnesota, USA.

Carlo Salvarani, MD;
Robert D. Brown Jr, MD;
John Huston III, MD;
Gene G. Hunder, MD.

Please address correspondence and reprint requests to: Carlo Salvarani, MD, Servizio di Reumatologia, Arcispedale S. Maria Nuova, V.le Risorgimento 80, 42100 Reggio Emilia, Italy.
E-mail: salvarani.carlo@asmn.re.it

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A 43-year-old woman with primary central nervous system vasculitis (PCNSV) experienced a relapse and presented with seizures. The diagnosis of PCNSV had been made at the age of 32 years by brain biopsy. Her initial clinical manifestations included headache, confusion, cognitive decline, personality change and drowsiness. At that time, an open brain biopsy showed granulomatous vasculitis. A cerebral angiography was normal. Treatment with oral prednisone and oral cyclophospahamide resulted in rapid improvement of her neurological status. However, she was unable to decrease her prednisone dosage to less than 20 mg/d because of a relapsing course.

At the time of the last relapse, magnetic resonance imaging (MRI) contrast enhanced T1 weighted axial (Fig. **A**) and coronal (Fig. **B**) sequences showed multiple zones of linear abnormal contrast enhancement in a perivascular distribution. These findings are compatible with vasculitis. MRI is very sensitive for PCNSV (1). MRI findings are abnormal in almost all the patients, however, the MRI appearance is usually not specific for CNS vasculitis. However, some investigators have suggested that the "perivascular" pattern of enhancement observed in our patient, although unusual, could be specific for PCNSV (2, 3). The observed changes may be an expression of vascular and perivascular inflammation that include meninges and surrounding gray and white matter.

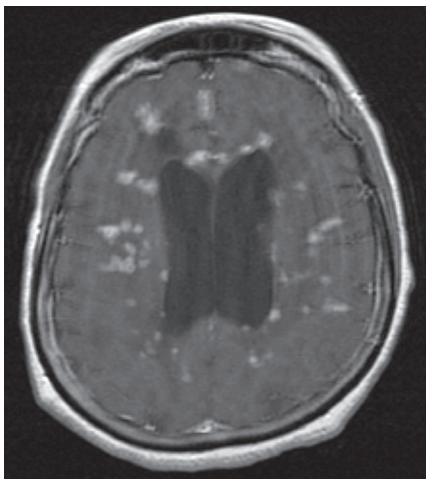

Fig. 1. A

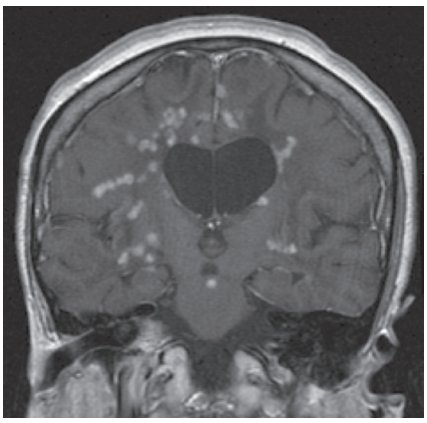

Fig. 1. B

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