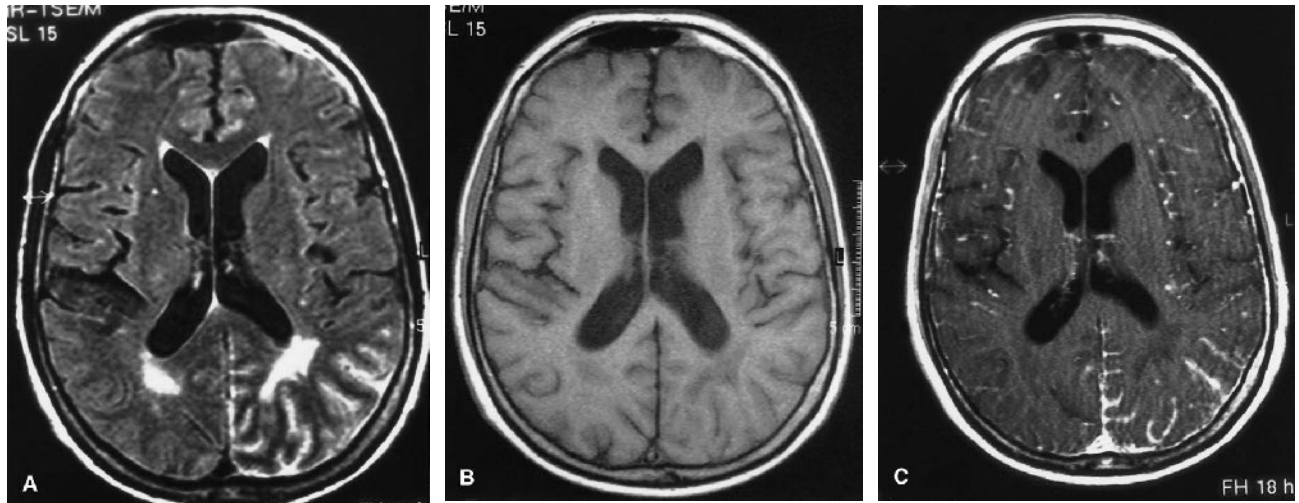


# Leptomeningeal Enhancement and Venous Abnormalities in Granulomatous Angiitis of the Central Nervous System

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**FIG. 1.** **A:** Axial FLAIR magnetic resonance imaging shows high signal in the subarachnoid space, most extensive over the left parietal lobe, as well as periventricular white matter chronic ischemic changes. **B:** Axial T1 unenhanced magnetic resonance imaging shows subtle low intensity areas in the peritrigonal white matter. **C:** Axial T1 enhanced magnetic resonance imaging shows diffuse leptomeningeal enhancement, most extensive over the left parietal lobe.

**Abstract:** A 68-year-old woman with a relatively acute onset of right homonymous hemianopia, Gerstmann syndrome, and global cognitive failure was found to have a lymphocytic pleocytosis and elevated protein on spinal fluid examination and displayed marked meningeal enhancement on magnetic resonance imaging and dilated cortical venules on cerebral angiography. Brain and meningeal biopsy disclosed a necrotizing granulomatous inflammation of small and medium-sized subarachnoid vessels. The brain parenchyma was normal. The angiographic presence of venous abnormalities, the lack of observable angiographic arterial involvement, and the lack of

parenchymal pathology are distinctly unusual in granulomatous angiitis of the central nervous system. This case, therefore, extends the pathologic and imaging spectrum of this disorder.

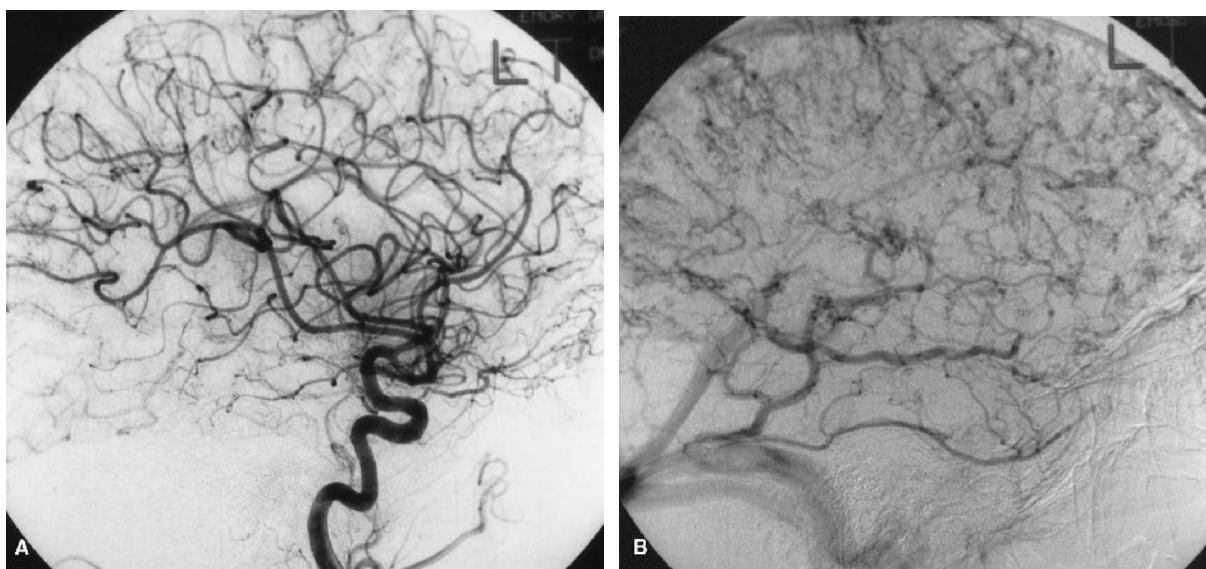
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A 68-year-old, right-handed, white woman was noted by her husband to be bumping into walls. Within 3 hours, she complained of right-sided visual impairment and pain behind her left ear. She seemed to have difficulty pronouncing words. Twenty-four hours later, when examined in the emergency room, the visual impairment and confusion persisted, but language skills had returned to normal. On examination, she recalled none of three words after a delay of 5 minutes. She also displayed agraphia, acalculia, right/left disorientation, finger agnosia, difficulty following complex commands, and a right homonymous hemianopia. Computed tomography (CT) of the head, performed without contrast, demonstrated periventricular white matter hypodensities consistent with small vessel ischemic disease.

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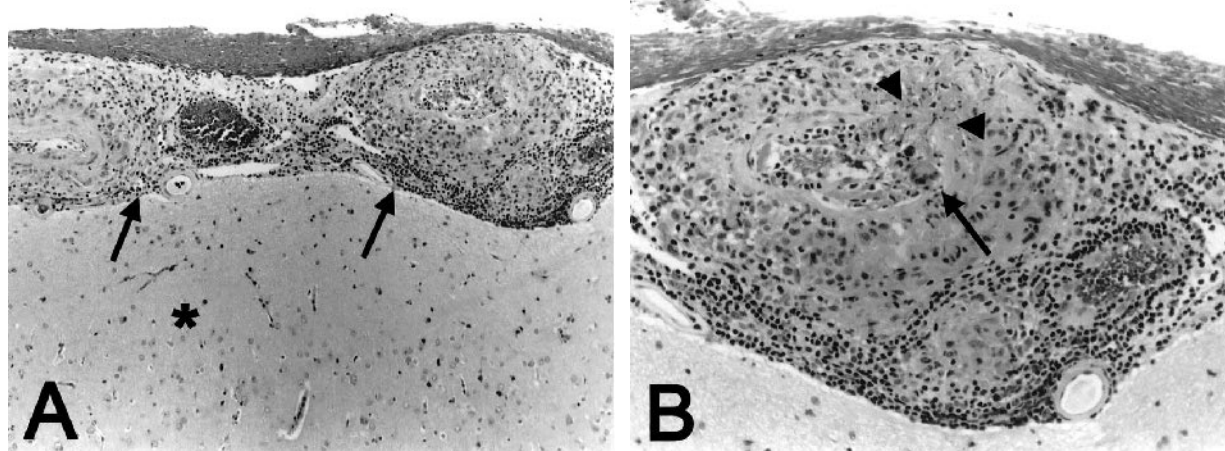


**FIG. 2.** Cerebral angiogram. **A:** Left anterior circulation, sagittal view, arterial phase. Normal arteries without evidence of vasculitis. **B:** Left anterior circulation, sagittal view, venous phase. Prominent, tortuous superficial cortical venules, most extensive over the parietal lobe. The venous sinuses are normal.

She was admitted to the neurology service and placed on intravenous heparin with the presumed diagnosis of acute cerebral ischemia. Despite therapeutic anticoagulation, she developed word finding difficulty and impaired comprehension. Magnetic resonance imaging (MRI) of the brain, performed 3 days after symptom onset, showed increased signal on FLAIR images in the convexity and sulcal spaces, most extensive over the left parietal lobe (Fig. 1A), and periventricular white matter chronic ischemic changes.

T1 unenhanced images show subtle low intensity areas in the peritrigonal white matter (Fig. 1B). Postgadolinium images showed extensive enhancement of the leptomeninges (Fig. 1C). Diffusion-weighted MRI failed to show restricted diffusion, ruling out acute ischemia. Magnetic resonance angiography (MRA) showed normal intracranial and extracranial vessels.

Heparin was stopped and three consecutive lumbar punctures were performed, each with elevated opening



**FIG. 3.** Meningeal and cortical biopsy. **A:** Subarachnoid space is widened by perivascular mixed inflammatory infiltrate (arrows). The underlying cerebral cortex (asterisk) is unaffected. **B:** Higher magnification of subarachnoid space shows a granulomatous, inflammatory infiltrate with epithelioid histiocytes and lymphocytes. There is a solitary multinucleated giant cell in a vascular wall (arrow). The histopathologic diagnosis of vasculitis was based on an active perivascular inflammatory infiltrate causing fibrinoid necrosis of the vascular walls (arrowheads).

pressure (300–350 mm water), elevated protein (106–223 mg/dl), lymphocytic pleocytosis (8–38 white blood cells), and negative cytology and cultures. A four-vessel cerebral angiogram showed normal arterial vasculature (Fig. 2A). However, in the venous phase, there were prominent and tortuous superficial cortical venules, most extensive over the left parietal region (Fig. 2B). Meningeal and cortical biopsy, obtained because of worsening mental status, revealed an active granulomatous, necrotizing inflammation and fibrinoid necrosis of small and medium-sized vessels in the subarachnoid space. There was no evidence of parenchymal involvement (Fig. 3). Based on the biopsy results, the patient was diagnosed with granulomatous angiitis of the central nervous system (GANS) and treated aggressively with high-dose prednisone and cyclophosphamide. She had dramatic improvement in her neurologic status within days.

Granulomatous angiitis of the central nervous system (GANS), also called primary angiitis of the central nervous system or primary central nervous system vasculitis, is an idiopathic inflammatory disorder of small central nervous system leptomeningeal and parenchymal blood vessels (1). The typical MRI findings in GANS are multiple, bilateral, T2 and FLAIR high signal abnormalities in the cortex and deep white matter. Even when conventional MRI is normal, signals consistent with ischemia may be demonstrated on diffusion-weighted MRI (4). Only once has leptomeningeal enhancement been reported as the only MRI abnormality (3). In that case (3), the patient had acute onset of confusion and gait impairment, diffuse leptomeningeal enhancement

on MRI, lymphocytic pleocytosis and elevated protein on cerebrospinal fluid examination, and a noninfectious granulomatous inflammation of subarachnoid vessels demonstrated on biopsy. However, that patient did not undergo cerebral angiography or diffusion-weighted imaging. Our patient's diffusion-weighted imaging was normal, confirming lack of brain parenchymal ischemia. Moreover, there was no evidence of arteritis on conventional angiography. In fact, cerebral angiography is only 60% sensitive in pathologically documented cases of GANS; the "classic" findings of arteritis—alternating areas of stenosis and ectasia in multiple vascular distributions—occur in less than 40% of cases (2). The extensive abnormalities seen on the venous phase of our patient's angiogram (Fig. 2B) are very unusual and appear to correlate with the leptomeningeal enhancement seen on MRI. This finding likely represents either venous inflammation or localized venous dilation secondary to the overlying leptomeningeal inflammation.

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