

Vasculitis Presenting as Primary Leptomeningeal Enhancement with Minimal Parenchymal Findings

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Summary: In this case of biopsy-proved granulomatous angiitis, MR showed prominent leptomeningeal enhancement with comparatively little parenchymal involvement. Pathologically, granulomatous angiitis chiefly affects small vessels, with a predilection for those in the leptomeninges. Although marked leptomeningeal enhancement is not the most common finding, vasculitis should be added to the differential diagnosis of this MR appearance.

Index terms: Angiitis; Vasculitis; Meninges, magnetic resonance

In general, when vasculitis is suspected, evidence of parenchymal ischemia is sought (1, 2). While computed tomography (CT) and magnetic resonance (MR) may show signs of infarction, the best modality for the diagnosis of vasculitis, short of biopsy, is cerebral angiography (3–8). In a patient without imaging findings of parenchymal ischemia and any characteristic angiographic changes, vasculitis involving the central nervous system would be considered unlikely; however, vasculitis can manifest primarily as striking leptomeningeal enhancement on contrast-enhanced MR, as shown by this case of biopsy-proved granulomatous angiitis of the brain (1–10).

Case Report

A 53-year-old woman was admitted with severe headache, confusion, and gait difficulties and was suspected of having a subarachnoid hemorrhage. All laboratory tests were normal, except for lumbar puncture, which revealed over 300 red blood cells per μL . The puncture was thought to be traumatic since 100 white blood cells, 80% lymphocytes, were also found. Two different CT scans and cerebral angiography were unremarkable.

Noncontrast MR imaging of the brain on the second hospital day showed a few small foci of increased signal intensity in the periventricular white matter bilaterally on long TR/TE images, consistent with nonspecific white

matter disease (Fig. 1). Repeat noncontrast MR 11 days after admission was unchanged, as was a repeat CT scan. Contrast-enhanced MR on the 22nd hospital day revealed diffuse leptomeningeal enhancement (Fig. 2). Long TR MR images showed possible new foci of increased intensity in the right insula and in the right medial occipital temporal gyri but were marred by motion artifact.

Multiple cerebrospinal fluid (CSF) studies revealed increased CSF pressure and protein, up to 550 mm and 1440 mg per 100 mL, respectively. Red blood cells decreased to under 60; white blood cells were elevated at 50 to 122, 85% of which were lymphocytes. No evidence of bacterial, viral, or fungal infection or tumor was found. The patient was treated with antibiotics for the provisional diagnosis of chronic meningitis of unknown etiology. Repeat MR 29 days after admission revealed multiple small bilateral cortical foci of increased signal intensity on short and long TR images, consistent with hemorrhagic changes. Additional areas of mild hypointensity on short TR images and hyperintensity on long TR images were also seen, suggestive of ischemia. On postcontrast MR, diffuse leptomeningeal enhancement was again seen as well as enhancing foci near the right sylvian fissure, most consistent with evolving



Fig. 1. On admission. Long TR (2951/80) image reveals small hyperintense lesions in the deep white matter.

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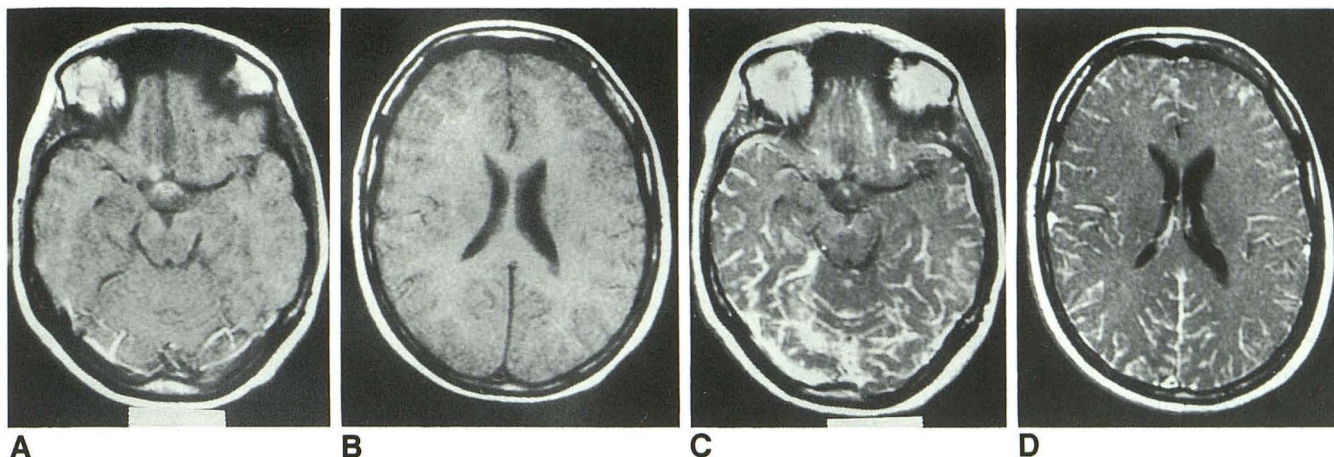


Fig. 2. Twenty-second hospital day.

A and B, Precontrast short TR (500/30) image with gradient moment nulling shows no definite abnormality.

C and D, Postcontrast short TR images (500/30) with gradient moment nulling disclose prominent leptomeningeal enhancement along the surface of the cortex. Although some vascular enhancement may be present, due to the addition of gradient moment nulling, the extent of enhancement is striking, invaginating into all of the sulci.

infarction (Fig. 3). An infectious cause was still suspected.

On the 30th hospital day, meningeal and brain biopsy was performed. At surgery, biopsy appeared unremarkable. Histologic examination, however, disclosed diffuse transmural granulomatous inflammatory infiltrates with multiple plump epithelioid cells, Langhans' giant cells, and chronic inflammatory cells in the vessels of the meninges and the cortex. Bacterial, fungal, and acid-fast-bacillus stains and cultures were negative. The pathologic diagnosis was granulomatous angiitis.

The patient received high dose methyl-prednisolone therapy (1 g per day for 5 days) followed by prednisone (100 mg daily) and showed gradual improvement. At discharge, she was fully alert and able to walk with assistance.

Discussion

Granulomatous angiitis of the brain is a vasculitis of the central nervous system of unknown origin. It is frequently fatal unless the patient receives adequate treatment with corticosteroids or immunosuppressives (3-9). The typical clinical symptoms are headache, mental status changes, focal signs and, occasionally, spinal cord involvement and seizure activity (3-7). CSF findings commonly reveal lymphocytic pleocytosis, elevated protein, and normal glucose. CT and MR classically show foci of ischemia or hemorrhage (3-10). Characteristic angiographic features in-

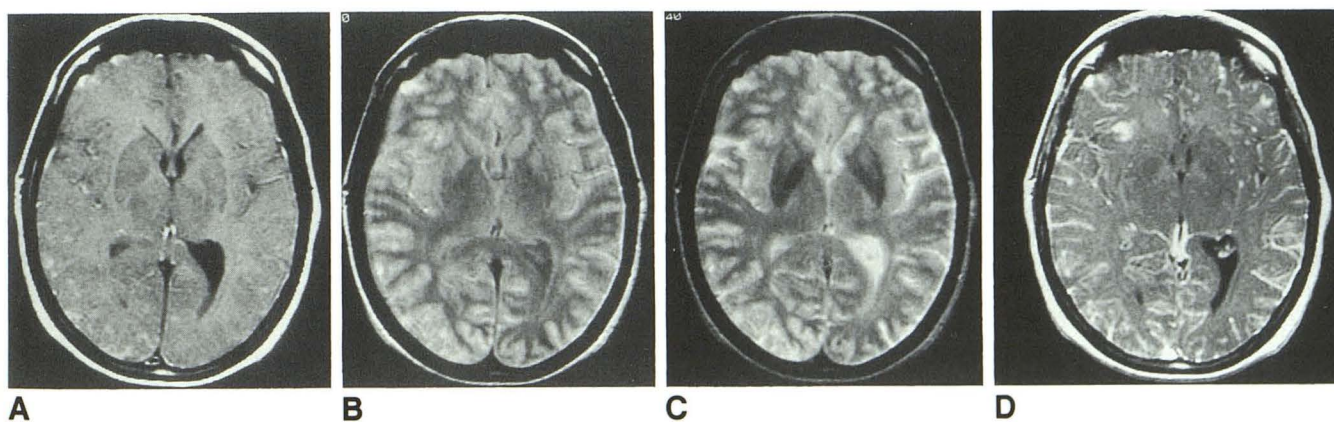


Fig. 3. Twenty-nine days after admission.

A, Short TR (500/30) scan with gradient moment nulling reveals multiple new small bilateral cortical foci of increased signal intensity, consistent with hemorrhage.

B and C, Long TR (2000/30, 80) scans show new lesions of increased intensity in the right frontal and occipital lobes.

D, Contrast-enhanced TR (500/30) scan with gradient moment nulling discloses diffuse marked leptomeningeal enhancement, as well as new foci of cortical enhancement, due to progressive ischemic changes.

clude segmental narrowing or "beading" of the affected arteries (3–12), although angiography was found to be negative in 29% of 48 cases, presumably because involvement is primarily of very small vessels (7). A biopsy specimen of the cerebral parenchyma and/or the leptomeninges is the only way to ascertain the diagnosis if other studies are negative (1–10). Other causes such as systemic inflammation or infection must be excluded.

Pathologically, diffuse involvement of the leptomeninges can be caused by many diseases. In the case of granulomatous angiitis, a special predilection for involvement of the small leptomeningeal vessels has been reported (1, 2, 3, 7, 8), differentiating this entity from granulomatous meningitis. There is segmental inflammation and necrosis of small leptomeningeal and parenchymal blood vessels, with surrounding areas of tissue ischemia or hemorrhage. The infiltrate is composed of mononuclear cells, multinucleated giant cells, granulomata, and plasma cells (1). Clearly affected are small arteries and veins with a diameter of less than 200 micrometers (5).

In conclusion, in this case of proved granulomatous angiitis, the MR findings were initially those of striking leptomeningeal enhancement with possible minor superficial cortical involvement of the brain. Subsequent MR studies confirmed the leptomeningeal enhancement and showed the progression to hemorrhagic and ischemic cortical changes; thus, central nervous

system vasculitis must be added to the differential list of those lesions that cause prominent meningeal enhancement on MR.

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