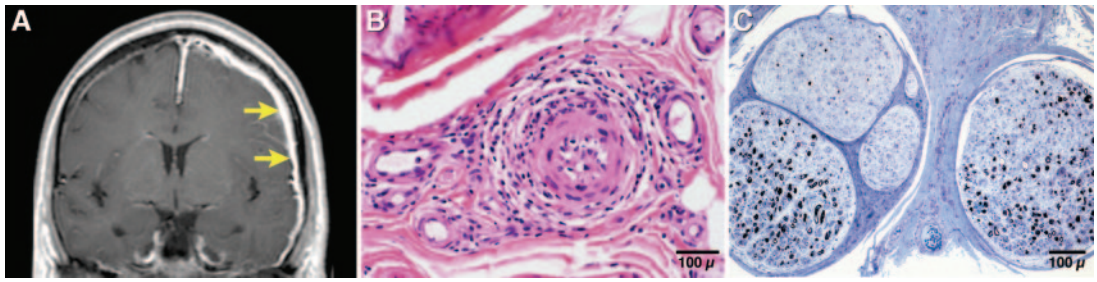
Hemi-meningitis

A focal sign heralding a multisystem necrotizing vasculitis

Figure Coronal brain MRI and sural nerve biopsy



(A) Coronal brain MRI with gadolinium demonstrating pachymeningeal enhancement (arrows). Sural nerve biopsy: (B) Paraffin cross-section shows a large epineurial arteriole with vessel wall damage, luminal occlusion, and recanalization. Inflammatory mononuclear cells are present around and within the vessel wall. (C) Methylene blue–stained, semi-thin epoxy section shows both intrafascicular and interfascicular multifocal fiber loss. The fascicles on the upper left are essentially devoid of myelinated fibers whereas those on the lower left and on the right retain numerous myelinated fibers, some actively degenerating.

A 58-year-old woman developed arthralgias and diffuse headache. Brain MRI with gadolinium revealed unilateral left pachymeningeal enhancement (figure, A) but CSF analysis revealed no evidence of infection or neoplasm. Four months later, she developed mononeuritis multiplex, nasal mucosal crusting, episcleritis, and digital splinter hemorrhages. c-ANCA autoantibody titer was elevated but chest X-ray and the serum creatinine level were normal. Nerve biopsy confirmed necrotizing vasculitis (figure, B and C). All clinical manifestations resolved after cyclophosphamide treatment.

The systemic features of this case are most consistent with Wegener granulomatosis, a necrotizing vasculitis with predilection for small vessels and diverse, often multifocal, neurologic manifestations.[1] The meninges receive lateralized vascular supply in parallel with the brain, including extensive branches from the internal carotid and vertebral arteries. The strikingly unilateral nature of the meningitis suggested a vascular etiology.

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1. Nishino H, Rubino FA, DeRemee RA, Swanson JW, Parisi JE. Neurological involvement in Wegener's granulomatosis: an analysis of 324 consecutive patients at the Mayo Clinic. Ann Neurol 1993;33:4–9.

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