

**Unilateral chronic relapsing primary central nervous system vasculitis**

Sirs,

Primary central nervous system vasculitis (PCNSV) is a rare disorder of unknown cause that is restricted to brain and spinal cord (1). Awareness of the wide spectrum of clinical presentations facilitates early recognition and treatment which may prevent serious outcomes or death. We report a case with chronic relapsing PCNSV involving only one cerebral hemisphere. This study was approved by the Mayo Clinic Institutional Review Board.

In November 2002 a 20-year-old previously healthy woman developed acute onset of headache, weakness of the left extremities, and left focal motor seizures with secondary generalisation. Spinal fluid (CSF) analysis showed increased protein (72 mg/dL); cultures/cytology were negative. Extensive blood tests including infectious disease-related serologies, autoantibody panel, and coagulation tests were negative or normal. Brain magnetic resonance imaging (MRI) showed multiple enhancing lesions involving the right cerebral hemisphere. MR angiography and conventional cerebral angiography were normal. A cerebral biopsy revealed lymphocytic vasculitis. Search for fungi, tubercle bacilli, and herpes simplex types 1 and 2 on the pathological specimens was negative. Immunocytological stains showed a benign vascular lymphocytic infiltration. Prednisone 60 mg/daily was started with 90% resolution of the left arm and leg weakness. In February 2003 prednisone was discontinued, neurologic symp-

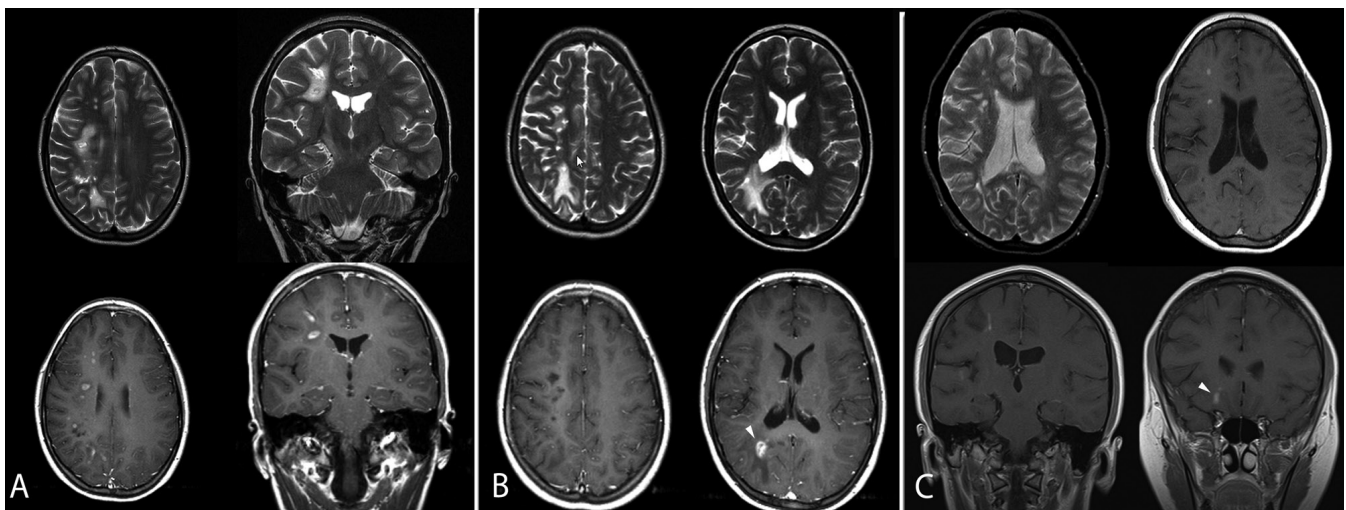
toms returned. Repeat MRI showed new areas of T2 hyperintensity and enhancement in the right hemisphere (Fig. 1A). The left hemisphere remained normal. A second biopsy in May 2003, while the patient was off prednisone confirmed the presence of an active lymphocytic vasculitis without evidence of demyelination. Prednisone 60 mg/day and oral cyclophosphamide (150 mg/day) were started. After 6 weeks, oral cyclophosphamide was suspended because of haemorrhagic cystitis. 4 months later mycophenolate mofetil 200 mg/day was begun, but leukopenia rapidly developed and it was stopped. In November 2004, she started azathioprine 200 mg/daily. In the following 6 years she continued the treatment with prednisone and azathioprine. However, she had several clinical exacerbations of the left-sided weakness and MRI changes (new areas of T2 hyperintensity and enhancement) (Fig. 1B) that coincided with attempts to lower the prednisone dose below 35–40 mg/day. In 2009, she was referred to Mayo Clinic. She was taking prednisone 40 mg/day and azathioprine 200 mg/day. Extensive blood tests were negative or normal. CSF showed slight increased protein (54 mg/dL), without oligoclonal bands and normal IgG synthesis rate and IgG index. Review of the previous biopsy specimens confirmed the presence of an active lymphocytic vasculitis only. MRI showed several small enhancing lesions involving the right hemisphere. Compared to an MRI performed 6 months before, there was some regression of enhancing lesions, but also small new lesions (Fig. 1C). Spine MRI was normal. Review of the several MRIs over the previous 6 years revealed the waxing and waning of the unilateral cerebral hemispheric pro-

cess since 2003, with new areas of T2 hyperintensity and enhancement appearing at different times and other lesions regressing (Fig. 1). The diagnosis of PCNSV was considered confirmed. A demyelinating process was excluded on the basis of its absence on the two brain biopsies. ADEM, Rasmussen's encephalitis, low-grade neoplasm or lymphoma, and infections were essentially excluded by the unilaterality, relapsing nature, long-term response to corticosteroid and immunosuppressive treatment, and brain biopsy findings. Intravenous cyclophosphamide at a dosage of 750 mg/meter squared/month was started. 6 months later she was able to halve the prednisone dose without relapses and with significant improvement of MRI lesions.

Abnormal MRI findings in PCNSV are characteristically bilateral and multiple (2, 3). Other than occasional cases presenting with a mass like lesion unilaterally involving a cerebral lobe (4), multifocal enhancing lesions confined at diagnosis to one cerebral hemisphere are rare (5). To our knowledge, this is the first case of adult PCNSV with multifocal relapsing disease confined to the same hemisphere during a long-term relapsing course.

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**Fig. 1.** Relapsing unilateral right cerebral hemisphere vasculitis over a 6-year time frame. **A.** 2003, Axial and Coronal T2WI (Top) demonstrating marked vasogenic oedema surrounding more focal ovoid T2 hyperintense lesions. Axial and Coronal Post gadolinium T1WI (Bottom) demonstrate ring enhancing, punctate and linear enhancing lesions in the right centrum semiovale. **B.** 3 years later, 2006. Axial T2WI (top) and post gadolinium T1WI (bottom) demonstrating resolution of the more superior enhancement and reduction in the mass like vasogenic oedema. Several new regions of vasogenic oedema and ring like enhancement in the right parietal occipital lobe have appeared (white arrowhead). **C.** 6 years later, 2009. Axial T2WI and post gadolinium T1WI (top) and coronal post gadolinium T1WI (bottom) demonstrated resolution of previous enhancement but several new regions of round and linear enhancement (white arrowhead) with focal T2 hyperintensities.

## Letters to the Editors

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