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Case report

Unihemispheric central nervous system vasculitis

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ABSTRACT

Patients with primary central nervous system vasculitis (PCNSV) usually manifest with multiple enhancing bilateral hemispheric lesions. We presented an extremely rare clinical course and follow-up of a patient with PCNSV affecting only a single (right) hemisphere. A 33-year-old previously healthy man presented with a left hand clonic seizure followed by a secondary generalized tonic-clonic seizure and dysarthria. MRI brain revealed multiple hyperintense lesions confined to only the right hemisphere with contrast enhancement, involving both white and grey matters. He was treated with a methylprednisolone for 5 days followed by prednisone for suspected acute disseminated encephalomyelitis without improvements. He was presented again with left-sided weakness, transient dysarthria and black objects in left visual field. MRI brain was unchanged. MR angiogram and conventional cerebral angiogram were normal. Autoimmune work-ups were all negative. A brain biopsy showed evidence of PCNSV. He was then successfully treated with intravenous cyclophosphamide followed by oral azathioprine. On a follow-up 3 years later, he remains asymptomatic on azathioprine and a repeat MRI showed all areas of enhancement were gone.

1. Introduction

Primary central nervous system vasculitis (PCNSV) is a rare, poorly understood neurological disorder and difficult-to-diagnose disease. The incidence is 2.4 cases per 1,000,000 person-years [1]. Most cases manifest with multiple enhancing bilateral hemispheric lesions. We describe the clinical course and follow up of a patient with PCNSV affecting only a single (right) hemisphere and review the literature on this very rare presentation.

2. Case

A 33-year-old man, who was in an excellent health, suddenly noticed twitching of his left hand followed by dysarthria, progressing to a generalized tonic-clonic seizure. Brain magnetic resonance imaging (MRI) with and without contrast revealed multiple hyperintense lesions on T2 involving the white matter as well as the cortex in the right hemisphere with contrast enhancement. There was no involvement of the left hemisphere, surrounding edema, or restriction on diffusion studies. He was treated with a 5-day course of methylprednisolone, followed by oral prednisone for suspected acute disseminated encephalomyelitis. Phenytoin was also initiated to control seizure. Three months later, he presented to our hospital with a 2-day history of left-sided weakness, transient dysarthria, and a transient black objects in his left visual field with associated frontal headache. A repeat MRI was unchanged from the previous MRI. MR angiogram and conventional cerebral angiogram were normal and did not show any evidence of vasculitis. Cerebrospinal fluid (CSF) analysis showed mild pleocytosis (9 white cells/uL (75% mononuclear cells, 25% lymphocytes), 4 red cells/uL) with total protein of 57 mg/dL and glucose 64 mg/dL. Other CSF studies were normal, including, IgG index, IgG synthesis rate, oligoclonal bands and electrophoresis. Rheumatologic work ups were all negative (anti-SM, anti-RNP, anti-SSA, anti-SSB, anti-SCL70, anti-centromere, anti-chromatin, anti-DsDNA, anti-JO1, anti-Ribosomal P, ANCA, and anti-Citrulline). C-reactive protein and erythrocyte sedimentation rate (ESR) were within normal limit. Hepatitis B and C profiles were negative. He was readmitted few weeks later after developing multiple partial motor seizures. Phenytoin was switched to levetiracetram which controlled his seizures. MRI showed enlarging and new lesions restricted to the right hemisphere with enhancement following gadolinium (Fig. 1A, B). A brain biopsy was eventually performed and revealed perivascular lymphocytic infiltrates, consistent with primary angiitis of the central nervous system (CNS) (Fig. 2). After a brain biopsy confirmed a PCNSV, he was started on oral prednisone 60 mg daily and intravenous cyclophosphamide 500 mg/m^2 and received 6 monthly infusions. He improved clinically with no residual weakness or seizures. A repeat MRI of the brain after completion of the 6 month course of intravenous cyclophosphamide showed dramatic improvement and most of the right hemispheric lesions had disappeared. He was switched to oral azathioprine 150 mg daily and

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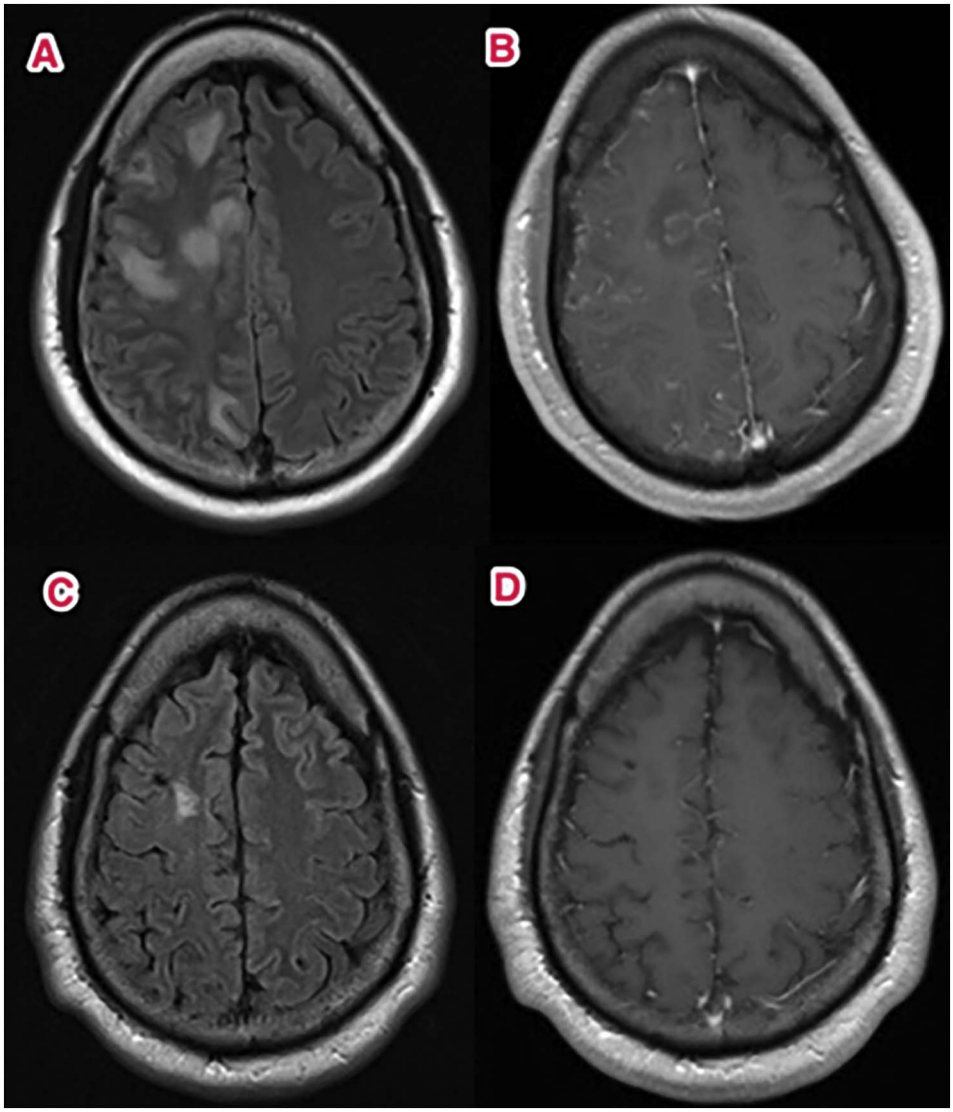

Fig. 1. MRI brain (FLAIR and T1 with contrast).
A: MRI brain (FLAIR) during the third admission before a biopsy showing abnormal right hemispheric patchy signal changes B: MRI brain (T1 with contrast) during the third admission before a biopsy showing multiple areas of contrast enhancement C: MRI brain (FLAIR) showing an improvement in size and number of previous hyperintense lesions after 3 years of treatment D: MRI brain (T1 with contrast) after 3 years of treatment showing no more enhancing lesions.

prednisone was tapered slowly and discontinued. A repeat MRI brain 6 months later showed a single residual enhancement, so azathioprine was increased to 200 mg daily. On a follow-up 3 years later, he remains asymptomatic on azathioprine 200 mg daily and levetiracetram 1500 mg twice a day and a repeat MRI (Fig. 1C, D) showed all areas of enhancement were gone.

3. Discussion

This case was extremely unusual and challenging diagnostically since the patient presented with multiple enhancing lesions, all of them remained confined to the right hemisphere. The initial diagnosis was challenging and the differential diagnosis included CNS lymphoma, multicentric glioma, multiple abscesses and Rasmussen encephalitis. The diagnosis was ultimately confirmed by brain biopsy and the patient responded very well to immuosuppressive therapy.

Calabrese and Mallek suggested the diagnostic criteria for diagnosing PCNSV which included the neurological deficits that cannot be explained by other etiologies and a cerebral angiography or central nervous system biopsy showing evidence of vasculitis [1,2]. A large cohort showed that the most common initial presentations are focal neurologic deficit and headache. Seizure is the first presentation in 16% in this series [1]. ESR (> 30 mm/h) may be uncommonly increased. CSF may show pleocytosis with white cells ranging from 0 to 535 cells/mL with median of 5 cells/mL, and elevated total protein concentration with median of 72 mg/dL. Cerebral angiography showed changes characteristic of vasculitis in 90%. Multiple-vessels abnormalities were abnormal in 93%. Although abnormalities on MRI brain are variable,