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Unilateral posterior reversible encephalopathy syndrome: A case report

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1. Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological entity that causes reversible subcortical vasogenic edema, predominantly in the parieto-occipital areas. The manifestations include headache, consciousness disturbance and encephalopathy [1]. Most cases show magnetic resonance imaging (MRI) abnormalities in the bilateral hemispheres, which are predominantly located in the posterior cortico-subcortical area [2]. Cases involving unilateral lesions are very rare. Furthermore, since PRES usually shows a reversible course, few reports include pathological data [2,3].

We herein report a case of pathologically confirmed unilateral PRES that showed repeated remission and recurrence.

2. Case report

A 73-year-old man was admitted to a local hospital complaining of right hemiparesis that had progressed over 2 days. His history included complete remission of gastric lymphoma (30 years previously), chronic hepatitis type C, hypertension, and chronic kidney disease. He had been treated with aspirin after a transient ischemic attack three years previously.

Brain MRI was performed on admission, and MR angiography (MRA) showed no vascular abnormalities (Fig. 1A). A high-intensity area was observed in the subcortical region of the left frontal-parietal region on fluid-attenuated inversion recovery (FLAIR) and apparent diffusion coefficient (ADC) mapping (Fig. 1B, C; white arrowheads), but diffusion-weighted imaging (DWI) showed almost isointensity to slight hyperintensity (data not shown). The lesion was also partly enhanced by gadolinium (Fig. 1D; yellow arrowheads). His initial blood pressure was 147/83 mmHg.

Although several neurological disorders, such as brain tumor, demyelinating disease, primary central nervous system (CNS) lymphoma, and progressive multifocal leukoencephalopathy (PML), were suspected based on the MRI findings, the clinical data were not sufficient to make a final diagnosis. Thus, after obtaining the patient's informed consent, a brain biopsy was performed at the left subcortical region of the parietal lobe two weeks after the onset of symptoms. A pathological analysis demonstrated no specific abnormalities, such as demyelination, excessive inflammation, or vasculitis, and only partial perivascular infiltration was observed (Fig. 2). Staining for CD3, CD5, CD20, and CD79a was also conducted for the differentiation of CNS lymphoma; however, no positive findings were obtained (data not shown).

His hemiparesis spontaneously improved, and he was discharged without any specific treatment. A second MRI scan performed one month later showed the attenuation of the unilateral white matter lesion in the left frontoparietal region; however, new white matter hyperintense lesions on both FLAIR and ADC mapping appeared in the bilateral temporal, occipital, and parietal subcortical areas (Fig. 1E, F; white arrowheads). He was therefore admitted to our hospital for a further examination.

On admission, he showed no focal neurological problems. A laboratory analysis revealed an increased blood urea nitrogen level of 32 mg/dl, a creatinine level of 2.26 mg/dl, and an sIL-2R level of 1107

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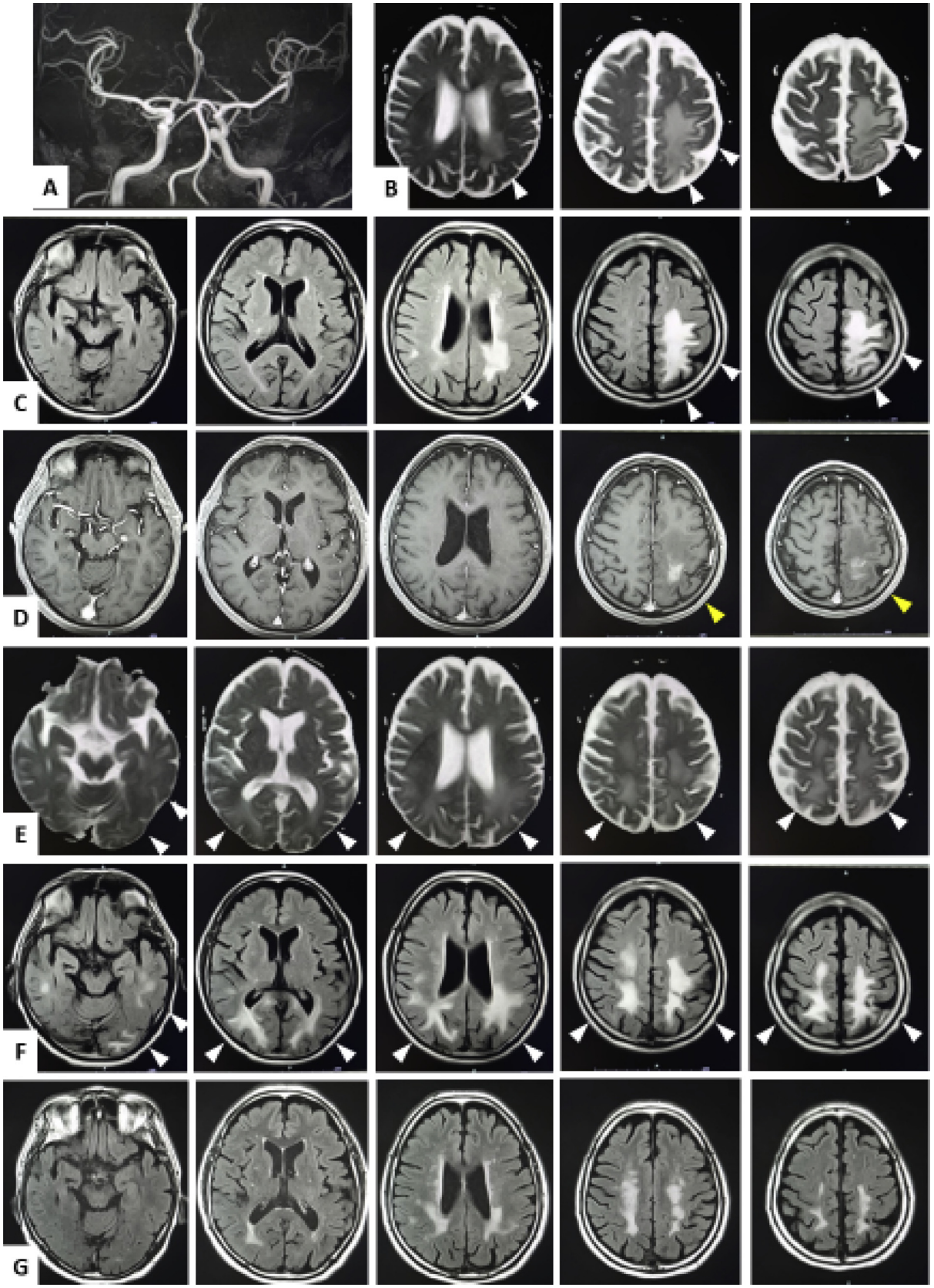

Fig. 1. Serial MRI images of the present case. MRA showed no vascular abnormalities (A). Initial brain MRI showed unilateral high intensity on both ADC mapping (B) and FLAIR (C) with partial enhancement with gadolinium (D; yellow arrowheads) in the left subcortical frontoparietal cortex (white arrowheads). A second MRI scan obtained one moths later showed improvement of the initial unilateral lesion but the appearance of a new subcortical lesion suggesting vasogenic edema on ADC mapping (E) and FLAIR (F) in the bilateral temporal, parietal, and occipital lobes (white arrowheads). A third MRI scan obtained three months later (G) showed improvement in the high-intensity area observed in series E and F (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article).

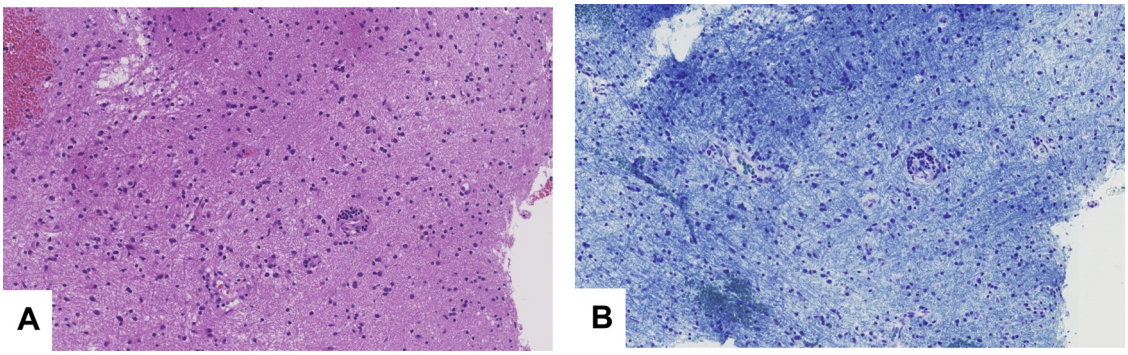

Fig. 2. Tissue stained with Hematoxylin and Eosin (H-E x20) showed only slight peri-vascular inflammation without excessive cell infiltration or tumor cells (A). Kluver-Barrera's (K-B x20) staining showed no evidence of demyelination (B).

U/ml. Other laboratory tests that might suggest collagen disease, vasculitis, or cancer were not conducted. A cerebrospinal fluid analysis revealed a normal cell count (6 cells/μl, 17% neutrophils, 83% lymphocytes), a protein level of 44 mg/dl, and a glucose concentration of 43 mg/dl. Thoracoabdominal computed tomography (CT) and abdominal echo revealed no evidence of cancer. However, the patient's blood pressure level was high, and 24-h ambulatory blood pressure monitoring indicated an average blood pressure of 167/89 mmHg. Surprisingly, the third MRI scan performed one month after the second one showed the spontaneous disappearance of the white matter lesion in both hemispheres (Fig. 1G).

We ultimately diagnosed this patient with PRES and strengthened his antihypertensive treatment. He was subsequently discharged without recurrence.

3. Discussion

PRES was reported as reversible posterior leukoencephalopathy syndrome in 1996 in patients with reversible white matter vasogenic edema in the posterior parieto-temporo-occipital region [1]. PRES was thought to be caused by vascular endothelial dysfunction due to several clinical conditions, such as rapid blood pressure fluctuation and the excessive production of cytokines in the vascular endothelium. The loss of the autoregulation of the cerebral blood flow and the impairment of the blood brain barrier can cause cerebral edema [2]. Bartynski et al. reported that MRI findings are classified into three types: "holohemispheric watershed pattern", "superior frontal sulcus pattern", and "dominant parietal occipital pattern", based on the main lesion distribution; other cases are classified as "partial or asymmetric expression of primary patterns", which is observed in 28% of all cases [4]. However, there are few reports of unilateral PRES.

We searched the PubMed database using keywords such as "PRES", "unilateral", and "asymmetric" and identified five cases of unilateral case of PRES among previous reports. All five cases were associated with intracranial vascular anomalies or conditions of vascular irritability (e.g. subarachnoid hemorrhaging or cerebral aneurysm). The analysis of each of the cases is provided in an attached Data in Brief article [5]. These vascular conditions might be related to the unilateral occurrence of the PRES. However, no obvious arterial abnormalities or vascular irritability were observed on MRI in our case. The present patient initially showed a unilateral subcortical white matter lesion mimicking a brain tumor or demyelinating disorders, but spontaneously improved, then the recurrence of bilateral white matter lesions was observed with a characteristic appearance of PRES.

There have been few reports on the pathological findings of PRES, and the previous five cases had not been pathologically examined. The reported pathological findings of PRES are largely nonspecific (e.g. mild edema or vacuolization of the white matter and mild nonspecific astrocytosis [without inflammatory findings or demyelination] to fibrinoid vascular necrosis) [2,3]. In the present case, brain tissue taken by brain biopsy, showed only slight peri-vascular inflammation without excessive cell infiltration or tumor cells, which were not specific findings for brain tumor, demyelination, or vasculitis. Thus we reported a case of PRES that showed unique but important clinical and radiological features.

4. Conclusion

The present findings suggest that PRES should even be considered in cases with a unilateral appearance, and careful follow-up with repeated imaging examinations should be implemented.

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