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Isolated cerebellar involvement in posterior reversible encephalopathy syndrome

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ABSTRACT

Background: Posterior reversible encephalopathy syndrome (PRES) is a serious and increasingly recognized disorder in humans. However, isolated cerebellar involvement in PRES is extremely uncommon. In this study, we sought to investigate its clinical and radiological features by describing a cohort of cases with PRES and isolated cerebellar involvement.
Methods: We report 2 patients with PRES with only cerebellar involvement and identified additional 9 cases using the PubMed database with the MeSH terms "posterior reversible encephalopathy syndrome", "hypertensive encephalopathy", "hypertension", "cerebellum", "encephalopathy", and "magnetic resonance imaging". We then collectively analyzed the clinical and imaging characteristics of these 11 cases.
Results: The average age was 28 years, with 8 male and 3 female patients. All cases had severe acute hypertension and T2 hyperintensity on MRI exclusively centered within the cerebellum. Of 11 patients, 7 had hypertensive retinopathy, a favorable clinical course with only antihypertensive treatment, and resolution of the cerebellar lesions on follow-up imaging. A total of 5 of the 11 patients received external ventricular drainage due to obstructive hydrocephalus and only 2 of the 11 had a seizure.
Conclusions: Isolated cerebellar involvement in PRES may be a unique variant that affects younger, male cases with severe acute hypertension and hypertensive retinopathy, but not necessarily seizure. Most patients have full recovery after fast control of blood pressure. Awareness of atypical neuroimaging features in PRES is critical for appropriate treatment.

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) is a reversible clinico-radiological syndrome that is clinically characterized by acute onset of headache, seizures, encephalopathy, and visual disturbances [1,2]. Radiologically, PRES is indicated by cortical and subcortical edema predominantly involving the parietal–temporal–occipital regions of the cerebral hemispheres [1,2]. Since 1996, PRES has become a heterogeneous syndrome caused by multiple factors [2,3]. The involvement of additional areas of the brain in PRES patients, including the frontal lobes, brainstem, basal ganglia and cerebellum, has also been reported [1–5]. However, to the best of our knowledge, isolated cerebellar involvement in PRES has been rarely reported [6–13] and little is known about its precipitating factors, course, radiological features, and outcome(s). In this study, we sought to widen the disease spectrum by describing a cohort of patients presenting with PRES and isolated cerebellar involvement.

2. Methods

We describe here 2 adult cases of PRES that presented only with cerebellar involvement. We searched the PubMed database using the Medical Subject Heading (MeSH) terms "cerebellum", "posterior reversible encephalopathy syndrome", "magnetic resonance imaging"; "hypertensive encephalopathy", "hypertension", and "encephalopathy". We included literature published in English describing cases with adequate clinical and radiological data to ascertain age, sex, symptoms, physical findings, neuroimaging, and clinical outcome. We considered clinical and radiological features as present if they were described and absent if they were not described.

2.1. Case 1

A 33-year-old man with a history of uncontrolled hypertension presented to an outside hospital with recurrent episodes of mild headache,

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ataxia, and nausea lasting for 10 days. The patient was neither immune-compromised nor septic. He was initially treated for hypertensive emergency (blood pressure 230/120 mm Hg), but required transfer to our institution three days later due to concerns of cerebellar infarction on head MRI. A chest examination revealed no abnormal findings. An abdominal sonographic study showed bilateral and chronically shrunken kidneys. There was no hepatomegaly or splenomegaly in the abdominal examination. Echocardiography revealed left ventricular hypertrophy. Laboratory results revealed high levels of creatinine (2.8 mg/dl). Results of blood tests, including a complete blood cell count, glucose, cholesterol, erythrocyte sedimentation rate, homocysteine, and C-reactive protein were normal. The results of serological tests for collagen disease as well as cerebrospinal fluid (CSF) testing were negative. The neurological examination revealed no specific findings. However, papilledema and hemorrhage were found upon funduscopic examination (Fig. 1A). Electroencephalographic results were unremarkable. Fluid-attenuated inversion recovery (FLAIR) images following presentation showed bilateral, hyperintense signal intensity changes in the cortex and subcortical white matter of the cerebellum (Fig. 1B). It should be noted that the parietal–temporal–occipital regions were not involved in FLAIR images (Fig. 1C). The susceptibility-weighted imaging (SWI) and T2*-weighted imaging were not available. However, diffusion-weighted imaging (DWI) did not reveal any abnormalities. Magnetic resonance angiography was also normal (Fig. 1D). There were no obvious abnormal enhancements in the affected areas after the injection of contrast material. Subsequently PRES causing vasogenic edema was considered as the likely etiology. We decided to aggressively control the patient's blood pressure. Thus, intravenous sodium nitroprusside (0.25 μg/kg/h) was used to control the patient's elevated blood pressure, after which the patient was successfully transitioned to oral administered agents (Valsartan and Amlodipine). Once his blood pressure was controlled (150/85 mm Hg), the patient's condition improved over the next 72 h. A follow-up brain MRI examination was performed 10 days after the episode and showed almost complete resolution of the cerebellar lesions (Fig. 1E). The patient was discharged on his twelfth day in hospital with no neurological deficits. A subsequent outpatient renal biopsy suggested IgA nephropathy (Fig. 1F, G). To date, the patient continues to take oral antihypertensive agents (Valsartan and Amlodipine) and his condition was followed up by a nephrologist.

2.2. Case 2

A 21-year-old male patient presented to our facility with a three month history of recurrent episodes of mild headache, dizziness, and nausea. His headache was characterized by dull, bilateral temporal pain and had increased in severity one week prior to his presentation. His medical history was unremarkable and he was neither immune-compromised nor septic. He had no vomiting, blurred vision, or limb weakness. All findings on his physical and neurological examination were normal. However, orbital funduscopy disclosed bilateral subretinal hemorrhage and papillary edema (Fig. 2A). On admission, his blood pressure and heart rate were 210/140 mm Hg and 120 bpm, respectively. Laboratory evaluations and CSF analysis were unremarkable. Head MRI revealed multiple, bilateral lesions in the cerebellum (Fig. 2B), but supratentorial structures were unremarkable on a subsequent FLAIR image (Fig. 2C). Neither SWI nor T2* imaging was available. Finally, there was no enhancement after the injection of contrast material (Fig. 2D). Based on our previous case study, we recognized that the lesions were likely secondary to a patient's hypertensive emergency. Blood pressure was subsequently and gradually controlled at 150/90 mm Hg over two days using intravenous nicardipine infusion followed by initiation of oral antihypertensives. His headache improved after 48 h of blood pressure control. Follow-up MRI images collected 14 days after presentation showed complete resolution of the previous diffuse hyperintensities at both cerebellar areas (Fig. 2E, F). The patient was subsequently discharged home with oral Lisinopril and has since been regularly monitored in our outpatient department. He has been in satisfactory and stable condition for more than one year.

3. Results

Eight reports of nine patients met the inclusion criteria [6–13]. These included our two case studies, resulting in a total of 11 cases for this descriptive analysis.

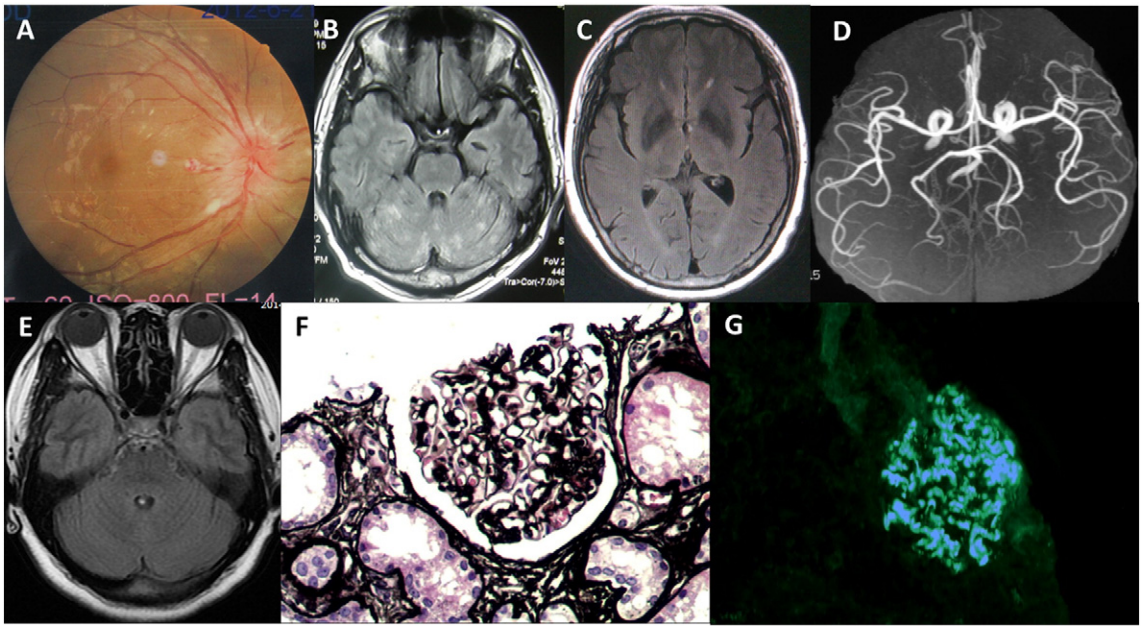

Fig. 1. Imaging findings in Case 1. A. Fundus photograph of the left eye shows optic nerve edema, arteriolar narrowing and hemorrhage. B–C. FLAIR MR images show hyperintense lesions restricted in the cortex and subcortical white matter of the cerebellum (B) but no abnormal hyperintensities outside of the cerebellum (C). D. Magnetic resonance angiography demonstrates no abnormalities. E. Follow-up MRI at 10 days demonstrates the complete resolution of the cerebellar lesions. F. Renal biopsy shows mesangial cell proliferation and focal thickening of the glomerular basement membrane (PASM staining, ×200). G. Immunofluorescence indicates positive IgA deposition in the mesangial area (×100).