

Acute chorea in posterior reversible encephalopathy syndrome

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Dear Sirs,

Posterior reversible encephalopathy syndrome (PRES) is a characteristic syndrome caused by reversible cerebral edema due to dysfunction of the cerebrovascular blood-brain barrier unit [2]. The major symptoms are clinical seizures, encephalopathy, visual symptoms, and headache, in combination with a typical brain magnetic resonance imaging (MRI) pattern of high signal intensity on T2-weighted images predominantly in the posterior regions [5]. After the first case series described as posterior reversible leukoencephalopathy in 1996 [3], numerous case reports, series and imaging studies have been published, including those with atypical features. However, to our knowledge, acute chorea as the presenting symptom of PRES has not been reported so far. We describe a patient with end-stage renal disease (ESRD) on hemodialysis who presented with acute chorea associated with bilateral basal ganglia lesions as an atypical feature of PRES.

A 50-year-old man visited to emergency department with an acute involuntary movement of “dance-like” feature after hemodialysis. He had a history of hypertension, diabetes and ESRD on dialysis for last 3 years (switched from peritoneal dialysis to hemodialysis 1 month ago). His initial blood pressure ranged from 178/91 to 210/110 mmHg, other vital signs were within normal range. He showed dysarthria and choreic movement of bilateral upper and lower extremities. He could not walk without assistance due to exaggerated steps with

a dance-like quality. The rest of neurological examination was normal. His initial laboratory data were clinically insignificant except elevated levels of blood urea nitrogen (29.0 mg/dL) and serum creatinine (6.11 mg/dL). Cerebrospinal fluid analysis was normal, excluding mildly elevated protein level of 59 mg/dL. Electroencephalography did not display epileptiform discharges nor ictal rhythm. Brain computed tomography revealed symmetric low attenuation of bilateral basal ganglia. Brain MRI demonstrated symmetric hyperintensity of bilateral basal ganglia, frontal and occipito-parietal lobes on fluid-attenuated inversion recovery (FLAIR), T2- and diffusion-weighted images with high apparent diffusion coefficient (ADC) values, suggesting vasogenic edema (Fig. 1a–d). Gadolinium-enhanced MRI was not performed because of advanced renal failure. Brain magnetic resonance angiography was unremarkable. His neurologic symptoms improved gradually after conservative management and blood pressure control (target range of systolic blood pressure of 140–150 mmHg and diastolic blood pressure of 80–90 mmHg). Follow-up MRI after 10 days of onset of symptom, showed markedly decreased high signal intensity of the lesions (Fig. 1e–h). He discharged with only subtle dysarthria without chorea after 2 weeks of admission.

Although there are reported cases of PRES with atypical neuroimaging features involving basal ganglia [5, 6], acute chorea as the presenting symptom with basal ganglia lesions in PRES has not been previously reported. Chorea is an irregular hyperkinetic involuntary movement originating from a dysfunction of neuronal networks interconnecting basal ganglia and the motor cortical area [1]. Among the various conditions related to chorea, there are rare case reports of acute chorea with bilateral basal ganglia lesions in patients with ESRD on dialysis, without an

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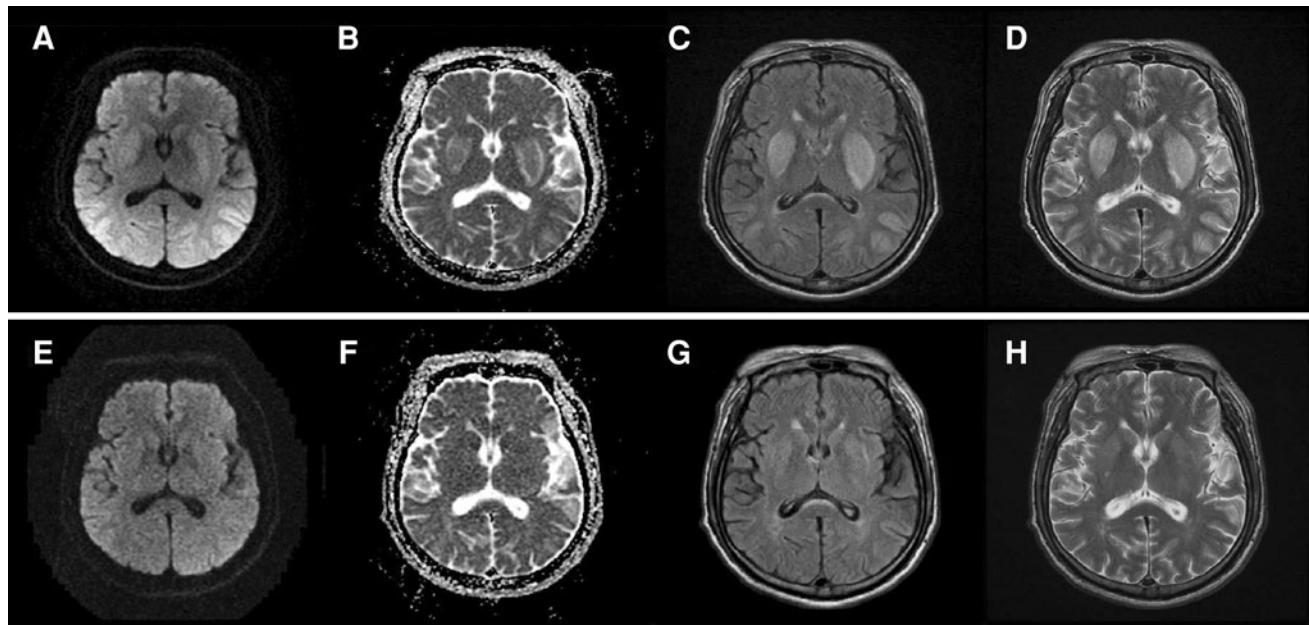


Fig. 1 Brain MRI at the onset of symptoms (a–d) and after 10 days (e–h). Initial MRI revealed bilateral symmetric diffusion-restriction in basal ganglia and parieto-occipital lobes on diffusion-weighted images (a) with high ADC values (b) suggesting vasogenic edema.

FLAIR (c) and T2-weighted images (d) showed high signal intensity of the same lesions. Follow-up MRI after 10 days displayed marked regression of the previous lesions (e–h)

understanding the pathophysiology [4, 7]. We thought our case was closer to PRES than these cases, because it showed not only basal ganglia lesions but also cortical lesions dominant in posterior regions which are characteristic of PRES. This case suggests that metabolic derangements related to dialysis together with hypertensive insult could cause reversible cerebral edema especially severe in basal ganglia. It might contribute to understand the possibly shared pathophysiologic spectrum of PRES and basal ganglia injury in dialysis patients with hypertension.

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Conflicts of interest We have no conflict of interest to declare.

Ethical standard All human studies must state that they have been approved by appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

References

- Cardoso F, Seppi K, Mair KJ, Wenning GK, Poewe W (2006) Seminar on choreas. *Lancet Neurol* 5:589–602
- Feske SK (2011) Posterior reversible encephalopathy syndrome: a review. *Semin Neurol* 31:202–215
- Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, Pessin MS, Lamy C, Mas JL, Caplan LR (1996) A reversible posterior leukoencephalopathy syndrome. *New Engl J Med* 334:494–500
- Kiryuk K, Khan F, Valeri A (2008) Acute chorea and bilateral basal ganglia lesions in a hemodialysis patient. *Kidney Int* 73:1087–1091
- Lee VH, Wijdicks EF, Manno EM, Rabenstein AA (2008) Clinical spectrum of reversible posterior leukoencephalopathy syndrome. *Arch Neurol* 65:205–210
- Liman TG, Bohner G, Heuschmann PU, Endres M, Siebert E (2012) The clinical and radiological spectrum of posterior reversible encephalopathy syndrome: the retrospective Berlin PRES study. *J Neurol* 259:155–164
- Yaltho TC, Schiess MC, Furr-Stimming E (2010) Acute bilateral basal ganglia lesions and chorea in a diabetic-uremic patient on dialysis. *Arch Neurol* 67:246