The Journal of Emergency Medicine, Vol. ■, No. ■, pp. 1–4, 2017

0736-4679/$ - see front matter

https://doi.org/10.1016/j.jemermed.2017.12.016

ELSEVIER

Clinical Communications: Pediatric

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME: A RARE CHILDHOOD CASE WITH UNCONSCIOUSNESS

Akira Komori, MD, Daisuke Mizu, MD, and Koichi Ariyoshi, MD, PHD

Kobe City Medical Center General Hospital, Hyogo, Japan

Reprint Address: Akira Komori, MD, Kobe City Medical Center General Hospital, 2-1-1 Minatojima Minamimachi, Chuo-ku, Kobe, Hyogo 650-0047 Japan

☐ **Abstract—Background: Posterior reversible encephalopathy syndrome (PRES) is a condition characterized by seizures, altered consciousness, visual disturbances, and headache. Characteristic findings on neuroimaging include cerebral edema, typically involving the parieto-occipital white matter. PRES has been associated with hypertension, autoimmune disease, and Henoch-Schölein purpura (HSP), but few cases have been reported, and fewer cases of PRES have been reported in children. Case Report: We report the case of a 4-year-old girl who presented with blindness and semi-consciousness. The patient had no significant medical history and no abnormalities on physical examination or laboratory testing, although she had slightly elevated blood pressure. After hospitalization, the patient showed some characteristic signs of HSP and cranial magnetic resonance imaging revealed PRES as the cause of semi-consciousness. In our discussion, we examine the clinical features of PRES and remarkable points for the clinical diagnosis and management of this rare but important disease. Why Should an Emergency Physician Be Aware of This?: Although reports of PRES in children are rare, PRES should be considered in the differential diagnosis of children presenting with disturbance of consciousness. Emergency physicians should consult with pediatric physicians to confirm diagnoses of PRES and determine an appropriate treatment plan, given its variable etiology. Measurements of blood pressure, which are often missing in pediatric cases, can help physicians to arrive at a correct diagnosis.** **© 2017 Elsevier Inc. All rights reserved.**

☐ **Keywords—childhood; hypertension; unconsciousness**

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a condition characterized by seizures, altered consciousness, visual disturbances, and headache. Characteristic neuroimaging features of PRES include cerebral edema, typically involving the parieto-occipital white matter. To date, few studies of PRES have been reported in children, and most of the scientific literature on PRES comprises case reports or small case series (1). Although the precise incidence of PRES in children is not known, it is thought to be rare. In one single-center pediatric intensive care unit study that included 2588 children, only 10 were diagnosed with PRES, resulting in an estimated incidence of about 0.4% (2).

CASE REPORT

Four days before visiting our hospital, a 4-year-old girl vomited a few times; however, based on her appearance and behavior, her mother did not believe she was ill. Her family doctor diagnosed her as having constipation when she visited him 2 days before visiting our hospital. She presented with sudden-onset eye irritation and loss of vision. Soon thereafter, she experienced consciousness disturbance and her mother called Emergency Medical Services. The child had no allergies, was not on any medication, and did not have any significant family history.

RECEIVED: 3 August 2017; FINAL SUBMISSION RECEIVED: 10 November 2017;
ACCEPTED: 1 December 2017

On clinical examination at the emergency department, the patient's consciousness level was 8 per the Pediatric Glasgow Coma Scale, and she was afebrile. She had no physical abnormalities or motor or sensory paralysis. Her heart rate was 145 beats/min, her blood pressure was 142/90 mm Hg, her respiratory rate was 28 breaths/min, and her peripheral capillary oxygen saturation was 100% under 6 L/min O_2. During her stay at the emergency department, she did not exhibit any seizures or convulsions. The patient's consciousness improved gradually without treatment after completion of a head computed tomography (CT) scan, which ended about 30 min after her arrival in the emergency department (score of 10 on the Pediatric Glasgow Coma Scale upon admission). The emergency department laboratory test findings are summarized in Table 1.

There were no remarkable abnormalities on blood testing, and urinary testing indicated ketone 3+ status, which may have been partly caused by her vomiting. Cerebrospinal fluid findings were negative for pleocytosis. We performed cranial CT, but did not detect any abnormalities (Figure 1). The patient was started on cefotaxime (75 mg/kg every 6 h) and acyclovir (20 mg/kg every 8 h) in consideration of the possibility of meningitis or herpes encephalitis, even though these diagnoses were unlikely based on the results of the cerebrospinal fluid examination. Oral nifedipine was administered to treat the patient's hypertension. At this time, the presence of secondary hypertension prompted us to consider the possibility of PRES.

The patient was hospitalized due to disturbance of consciousness, but showed improvements in consciousness and vision after the initiation of antihypertensive therapy. The next evening, she vomited once and complained of abdominal pain. On the third day of hospitalization, abdominal ultrasonography revealed swelling in the walls of the duodenum that was characteristic of Henoch-Schölein purpura (HSP) (Figure 2). At this time, diffuse palpable but painless purpura appeared on the patient's back. Based on these findings, we suspected a diagnosis of HSP and initiated methylprednisolone therapy (2 mg/kg/d). T2-weighted and fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) performed on the fourth day of hospitalization revealed high-intensity signal in the bilateral occipital lobes and gyral swelling that was more prominent in the left hemisphere (Figure 3). There was no diffusion restriction on diffusion-weighted imaging. These findings suggested angioedema, which was compatible with a diagnosis of PRES. At this time, purpura had extended to affect both of the patient's upper and lower legs, with new

Table 1. Emergency Department Laboratory Test Results

Laboratory Test	Value	Normal Range
Chemo		
CRP, mg/dL	2.49	<0.03
T-Bil, mg/dL	0.2	0.2–1.2
AST, IU/L	30	8–40
ALT, IU/L	8	8–40
LD, IU/L	264	120–250
Cr, mg/dL	0.34	0.40–0.80
BUN, mg/dL	10.3	8.0–20.0
Na, mEq/L	133	136–148
K, mEq/L	3.7	3.5–5.3
Ca, mg/dL	8.9	8.0–10.0
CBC		
RBC, $\times 10^6/\mu$L	494	350–510
Hb, g/dL	11.8	11.1–15.1
Ht, %	35.9	33.5–45.1
WBC, $\times 10^3/\mu$L	16.2	3.9–9.8
PLT, $\times 10^4/\mu$L	36.9	13.0–37.0
Neu, %	50.7	30.0–70.0
Lym, %	30.6	19.0–61.0
Mon, %	5.9	2.0–12.0
Eos, %	12.7	0–8.0
Urine		
pH	6.0	5.0–7.5
Glucose	(–)	(–)
Protein	(±)	(–)
WBCs	(–)	(–)
RBCs	(–)	(–)
Ketones	(3+)	(–)
CSF		
Cell count, /μL	4	0–4
Protein, mg/dL	14	15–45
Glucose, mg/dL	72	50–80

ALT = alanine transaminase; AST = aspartate aminotransferase; BUN = blood urea nitrogen; CBC = complete blood count; Cr = creatinine; CRP = C-reactive protein; CSF = cerebrospinal fluid; Eos = eosinophils; Hb = hemoglobin; Ht = hematocrit; LD = lactate dehydrogenase; Lym = lymphocytes; Mon = monocytes; Neu = neutrophils; PLT = platelet; RBC = red blood cell; T-Bil = t-bilirubin; WBC = white blood cell.

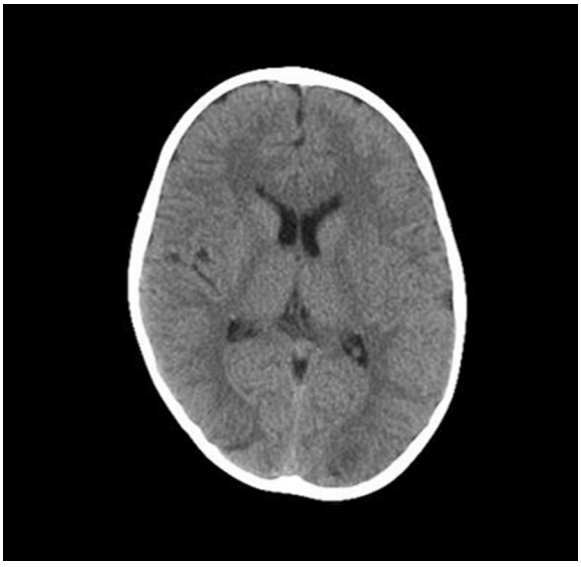

Figure 1. Head computed tomography scan on the day of admission.