



Clinical Notes

Immunoglobulin A vasculitis complicated with posterior reversible encephalopathy syndrome and reversible cerebral vasoconstriction syndrome

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Immunoglobulin A vasculitis (IgAV) involves vasculitis of the small vessels with deposition of immune complexes (IC) containing immunoglobulin A (IgA). Posterior reversible encephalopathy syndrome (PRES) is a rare neurological complication of IgAV, and presents with headache, seizures, and visual changes associated with reversible vasogenic edema. PRES can also occasionally occur with reversible cerebral vasoconstriction syndrome (RCVS) in adults. The presentation of both PRES and RCVS in children, however, is very rare. Herein, we report a case of IgAV with both PRES and RCVS in childhood.

A healthy 8-year-old Japanese boy presented with a 4-day history of acute abdominal pain. He was admitted to another hospital and antibiotic treatment was started. He suddenly had headache, a convulsion and consciousness disturbance on the day after admission. At 5 days after admission, based on elevated serum lipase and pancreas swelling on abdominal computed tomography, he was diagnosed with acute pancreatitis and was transferred to the present hospital for further evaluation and treatment. Physical examination on admission indicated a blood pressure (BP) of 120/72 mmHg, Glasgow coma scale score E3V3M6, and rebound abdominal tenderness. Serum amylase was 474 IU/L, lipase 361 U/L, and C-reactive protein, 3.86 mg/dL. I.v. meropenem was initiated. Brain magnetic resonance imaging (MRI) on the fifth day of hospitalization (Fig. 1a) showed hyperintense regions on T2-weighted, fluid-attenuated inversion recovery (FLAIR), and apparent diffusion coefficient (ADC) sequences in the right occipital lobe, indicative of PRES. Magnetic resonance angiography (MRA) showed multiple segmental narrowing in the right posterior cerebral artery (Fig. 1d). Purpura appeared on the ear and legs on the same day. Cerebrospinal fluid analysis on the eighth

hospital day was normal. Skin biopsy of the lesion indicated deposition of IC containing IgA. Thus, the diagnosis was IgAV. Factor XIII was also decreased. Meropenem treatment was stopped as the abdominal pain, serum amylase, and C-reactive protein gradually improved. Consciousness improved gradually and normalized on the 13th day of hospitalization. Follow-up MRI/MRA on the 13th hospital day showed improvement of the right occipital lobe hyperintense lesion on T2, FLAIR, and ADC sequences (Fig. 1b). Posterior cerebral artery narrowing disappeared completely (Fig. 1e). The clinical findings met the criteria of RCVS.¹ Hemorrhagic infarct was found in the right occipital lobe on MRI but was not treated because the visual disturbance was negative. The purpura disappeared on the 23rd hospital day. The patient was discharged on the 32nd hospital day without neurological defect. MRI and MRA were normal at 76 days after discharge (Fig. 1c,f). Informed consent for submission of the case report was obtained from the parents of the patient.

Disturbed regulation of cerebral arterial tone is a common pathological mechanism in PRES and RCVS. PRES is complicated in 17–38% of patients with RCVS.² PRES is a rare complication of IgAV. To our best knowledge, however, there are no reports of IgAV complicated with PRES and RCVS. Hypertension is a known cause of PRES, but a role for hypertension in PRES onset in the current case is unclear because BP was not measured at the previous hospital. Cerebral vasculitis secondary to IgAV can also cause blood–brain barrier breakdown and loss of autoregulation, resulting in PRES.³ Further, IC deposition in pancreatic vessels can cause pancreatic edema and small vessel embolisms, resulting in pancreatic inflammation.⁴ Thus, in the present case, IgAV may have led to PRES, RCVS, and acute pancreatitis.

The mean age of RCVS onset is approximately 42 years.⁵ Childhood RCVS is very rare, with 12 pediatric cases reported, only three of which were accompanied by PRES. These cases involved a 9-year-old boy with Loey–Dietz syndrome in which RCVS and PRES was triggered by operation,

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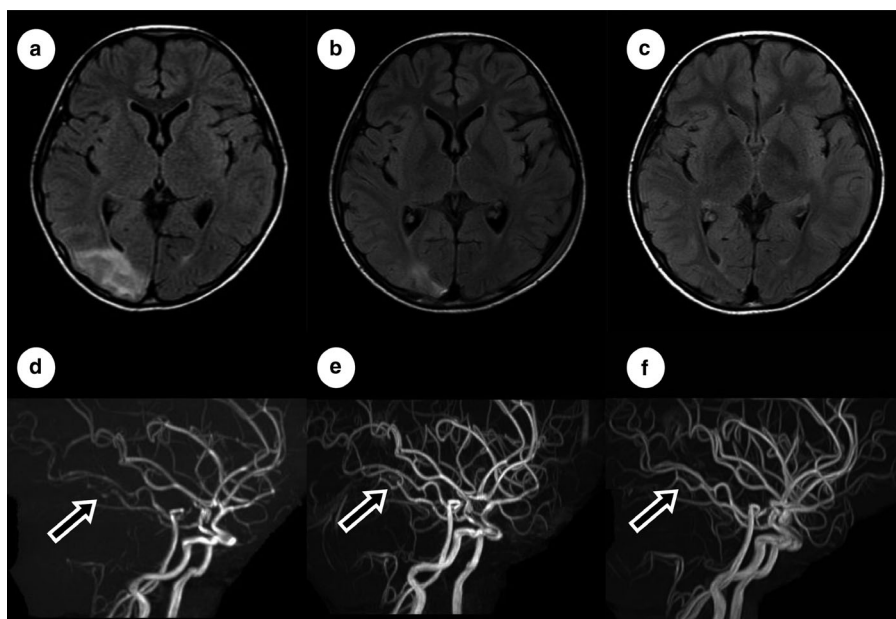


Fig. 1 (a–c) Brain fluid-attenuated inversion recovery magnetic resonance imaging and (d–f) magnetic resonance angiography on (a,d) the fifth hospital day, (b,e) 13th hospital day, and (c,f) approximately 2 months after discharge. Arrow, right posterior cerebral artery.

a healthy 13-year-old boy with RCVS and PRES triggered by exercise, and a 13-year-old boy with RCVS and PRES triggered by hemolytic uremic syndrome. Herein, we provide a new case of IgAV with both PRES and RCVS. Nevertheless, RCVS may be underestimated because MRA is not routinely evaluated in IgAV patients with PRES. Indeed, there are few reports of IgAV cases complicated with PRES that use MRA. Given that ischemic infarctions, subarachnoid hemorrhage, severe deficits, and death occur in 39%, 34%, 9%, and 2%, respectively, of RCVS patients,⁵ MRA should be performed in PRES cases.

In conclusion, pediatric cases of both PRES and RCVS are very rare. Both PRES and RCVS, however, can accompany IgAV, the most common vasculitis in childhood. MRA should be considered in IgAV patients with PRES.

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Disclosure

The authors declare no conflict of interest.

Author contributions

H.N. was involved in the clinical management of the patient, collected clinical information, and wrote the manuscript. K.K., T.K., and N.K. were involved in the clinical management of the patient. T.F. reviewed the manuscript. All authors read and approved the final manuscript.

References

- 1 Calabrese LH, Dodick DW, Schwedt TJ *et al.* Narrative review: Reversible cerebral vasoconstriction syndromes. *Ann. Intern. Med.* 2007; **146**: 34–44.
- 2 Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome; clinical and radiological manifestations, pathophysiology, and outstanding questions. *Lancet Neurol.* 2015; **14**: 914–25.
- 3 Woolfenden AR, Hukin J, Poskitt KJ *et al.* Encephalopathy complicating Henoch-Schönlein purpura: Reversible MRI changes. *Pediatr. Neurol.* 1998; **19**: 74–7.
- 4 Zhang Q, Guo Q, Gui M *et al.* Henoch-Schönlein purpura with acute pancreatitis: Analysis of 13 cases. *BMC Pediatr.* 2018; **18**: 159.
- 5 Singhal AB, Haji-Ali RA, Topcuoglu MA *et al.* Reversible cerebral vasoconstriction syndromes; analysis of 139 cases. *Arch. Neurol.* 2011; **68**: 1005–12.