Case Report

Rapid intracranial pressure drop as a cause for posterior reversible encephalopathy syndrome: Two case reports

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Abstract

Background: Posterior reversible encephalopathy syndrome (PRES) is characterized by reversible edematous lesions on radiological examinations as well as symptoms of altered consciousness and seizures. To date, the underlying mechanism remains largely unknown.

Case Descriptions: Case 1 is a 72-year-old man with a history of hypertension presented with a subarachnoid hemorrhage. Fourteen days after the successful clipping of a ruptured aneurysm; he experienced inadvertent overdrainage via the intraventricular drain. Nine hours later, he started to have seizures followed by disturbances in consciousness. An emergency magnetic resonance imaging showed multiple high-intensity lesions in the frontal, temporal, parietal, and occipital lobes, basal ganglia, brainstem, and cerebellar hemispheres bilaterally, which are compatible with typical magnetic resonance findings in PRES patients. He was treated conservatively and recovered well. Case 2 is a 68-year-old woman with a mild history of hypertension and a ventriculo-peritoneal shunt for obstructive hydrocephalus, who underwent a cysto-peritoneal shunt placement because of an enlarging symptomatic arachnoid cyst. Immediately following surgery, she experienced disturbances in consciousness and developed status epilepticus. Radiological examinations revealed remarkable shrinkage of the arachnoid cyst and multiple edematous lesions, which led us to strongly suspect PRES. With conservative treatment, her symptoms and the radiological abnormalities disappeared.

Conclusion: Based on the previous literature and our cases, we believe that the association between rapid reduction of intracranial pressure (ICP) and the development of PRES should be recognized because most neurosurgical procedures such as craniotomy or cerebrospinal fluid diversion present a potential risk of rapid reduction of ICP.

Key Words: Cerebrospinal fluid, intracranial pressure, lumbar puncture, posterior reversible encephalopathy syndrome

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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) was proposed by Hinchey et al. in 1996 to describe characteristic radiological changes indicating brain edema, especially distributed in the posterior circulatory area, which is the most apparent on fluid-attenuated inversion recovery (FLAIR) images.[14] These changes are also accompanied by symptoms, including altered mental status, seizure, cortical blindness and other visual abnormalities, headache, nausea, and vomiting.[14] These clinical and radiological presentations usually resolve within several weeks.

As the condition gained recognition, additional characteristics have been reported.[9] For example, radiological changes are not necessarily limited to the posterior lobes. Also, some irreversible symptoms may occasionally lead to death. In addition, we postulate an association between PRES and hypertension, immunosuppressive drugs, chemotherapy, renal insufficiency, autoimmune disease, eclampsia, preeclampsia, and sepsis. There is currently no single theory that can account for the mechanism of this condition.

Here, we present two patients who developed PRES immediately after a rapid reduction in intracranial pressure (ICP). We also reviewed the previous literature and propose that a reduction in ICP may be associated with PRES.

CASE PRESENTATIONS

Case 1
A 72-year-old man on regular medication for hypertension was transported to our hospital complaining of sudden headache followed by disturbed consciousness. Upon arrival, he showed an altered mental status with decreased alertness. He could follow commands but could not speak. He had no other focal deficits, such as hemiparesis or cranial nerve palsy. Computed tomography (CT) showed a subarachnoid hemorrhage (SAH) [Figure 1a]. Emergency angiography revealed a small aneurysm at the bifurcation of the left internal carotid artery and the posterior communicating artery [Figure 1b]. The aneurysm was clipped successfully on the day of admission, and his neurological conditions showed remarkable improvement. Because the angiogram obtained on day 6 showed a sign of localized mild spasm in the A1 portion of the left anterior cerebral artery, hypervolemic and permissive hypertensive therapy with a continuous ventricular drainage to control the ICP was implemented to prevent ischemic complications. His neurological status remained stable with no findings of ischemia on serial CT examinations; however, he became restless over time and started becoming agitated, likely because of the long period of bed rest. His blood pressure (BP) hovered around 170–190 over 80–100 mmHg. On day 14, he repeatedly got up and lied down on the bed, which caused an inadvertent overdrainage of cerebrospinal fluid (CSF) via the intraventricular drain. The amount of CSF drainage was as high as approximately 200 ml in

Figure 1: (a) Computed tomography (CT) scan upon admission showing a hematoma in the basal cistern. (b) Anteroposterior view of the left internal carotid angiogram revealing an aneurysm at the bifurcation of the internal carotid artery and the posterior communicating artery. (c) CT on day 14 showing ventricular narrowing and low-density area on the left occipital lobe (arrowhead). (d-f) Magnetic resonance (MR) imaging showed fluid-attenuated inversion recovery (FLAIR) images depicting diffuse high-intensity lesions in the bilateral frontal, temporal, parietal, and occipital lobes, and basal ganglia, brainstem, and cerebellum. (g) MR angiography showing no significant vasospasm. (h) MR imaging obtained on day 30 showing a complete resolution of high-intensity lesions.
2 h. His BP was 199/91 mmHg and no new neurological deficit was observed at this time. Emergency CT obtained 6 h after this episode revealed a ventricular narrowing compared to the previous CT images obtained before 2 days; in addition, a new, small, low-density area in the left occipital lobe was observed [Figure 1c]. Three hours after the CT examination, the patient had a tonic-clonic seizure followed by coma. Magnetic resonance (MR) images taken 9 h after the overdrainage showed diffuse hyper-intensity areas on FLAIR in the bilateral frontal, temporal, parietal, and occipital lobes, and basal ganglia, brainstem, and cerebellum [Figure 1d-f] without an apparent spasm on MR angiography [Figure 1g]. Based on the radiological findings, we made a diagnosis of PRES and started him on anticonvulsant and antihypertensive therapies. His neurological status made a slow but steady recovery over the following 9 days. MR imaging performed on day 30 demonstrated a complete disappearance of edema delineated as FLAIR high lesions [Figure 1h]. After ventriculo-peritoneal (VP) shunt placement for hydrocephalus and 3 months of rehabilitation, the patient was discharged home with no neurological deficits.

Case 2
A 68-year-old woman presented at our outpatient clinic complaining of left hemiparesis. She had a history of well-controlled hypertension and had a VP shunt placed 18 years ago for obstructive hydrocephalus due to midbrain cavernous malformation. CT images showed an enlargement of the right frontotemporal arachnoid cyst [Figure 2a], which we considered the cause of her left hemiparesis. A cysto-peritoneal shunt placement using a y-shaped connector was performed. Although there were no hemodynamic changes (her BP remained around 100/40 mmHg) or complications during the operation, the patient did not wake up after the anesthesia was completely terminated. Before the surgery, we had increased the valve pressure to 20 cmH₂O because we were concerned about a rapid reduction in ICP; however, postoperative CT demonstrated remarkable shrinkage of the cyst with a small amount of bleeding [Figure 2b]. Following surgery, the patient experienced tonic-clonic seizures and developed status epilepticus on the following day. The emergency CT images showed low-density areas in the bilateral occipital lobes [Figure 2c], and MR imaging on the fourth day following surgery revealed diffuse hyper-intensity areas on FLAIR imaging in the temporal, parietal, and occipital lobes on both sides, and the right frontal lobe [Figure 2d]. Because we strongly suspected PRES, we initiated anticonvulsant therapy. Although the patient had been comatose and unresponsive for >10 days, she gradually started to respond 12 days after surgery and showed neurological improvement. MR images taken 27 days after surgery demonstrated an almost complete disappearance of edematous areas [Figure 2e]. The patient required rehabilitation for another few months, but recovered to a normal status in 6 months.

DISCUSSION
Despite its name containing the term “posterior,” PRES is believed to occur in the posterior circulatory areas as well as in the temporal lobes or frontal lobes, as is also seen in the cases described here. PRES is associated with a variety of clinical conditions, including hypertensive status, immunosuppression, chemotherapy, renal failure, autoimmune disease, eclampsia, preeclampsia, and sepsis. Among these, previous studies have emphasized chronic hypertensive status as a predisposing factor for PRES because it is thought to injure the autoregulation of cerebral vessels and cause vasogenic edema. Although there was a history of hypertension in Case 2, the patient’s condition was mild and well controlled; Case 1 had high BP because he was in a state of post-SAH vasospasm, which could have induced vascular autoregulation dysfunction. It is well known that PRES can occur during SAH treatment. In 12 reports of 15 cases of PRES in patients with SAH reviewed, permissive or intensive hypertension employed during the acute phase of SAH to prevent the symptomatic vasospasm was presumed to be a cause of PRES. However, ICP was not directly measured, a rapid reduction in ICP appeared to occur in both of our cases. In Case 1, radiological clinical signs of PRES presented approximately 6 h after the inadvertent massive CSF drainage, indicating that the rapid decrease in ICP...
could have been the main trigger of PRES. Similarly, the postoperative CT in Case 2 showed substantial shrinkage of the cyst, suggesting a possible abrupt event that decreased ICP during surgery. Ho et al. proposed that reduction in ICP decreases ventricular size, resulting in mechanical stress to vessels and causing vasoconstriction and vasogenic edema.\(^{[15]}\) Some reports that measured ICP during the clinical manifestations of PRES found that the values were all within the normal range,\(^{[4,7,15,28]}\) possibly because of the timing of ICP measurement; autoregulation system dysregulation may recover quickly enough to adjust ICP values within a few hours before clinical symptoms become evident.

Hammad et al. recently reported a case of PRES secondary to CSF leak and intracranial hypotension, and also reviewed 10 cases of PRES that developed after spinal or epidural tap.\(^{[13]}\) They speculated that increased cerebral perfusion pressure due to an elevation of mean arterial pressure or a decrease in ICP resulted in hyperperfusion followed by vasogenic edema. On the basis of this proposal, we further expanded the literature search for reports on possible reductions of ICP because of invasive treatment or surgical procedure. A summary of 19 cases, including 10 cases in Hammad’s series, is shown in Table 1. Twelve cases were associated with a spinal tap or epidural anesthesia.\(^{[6,12,13,15,16,22,25,27,29,31–34]}\) Although not

| Authors and Year | Age and Sex | Past medical history | Related factors/procedures | Timing | Lesions | Outcome |
|------------------|-------------|----------------------|-----------------------------|--------|---------|---------|
| Torrillo et al. 2007 | 32 F | Nothing | Preeclampsia, labor, PDPH/epidural catheter | POD 4 | Bilateral F, P, O, ganglia, vermis | Full recovery |
| Hong et al. 2007 | 29 F | Nothing | Postural headache/CS, spinal anesthesia, blood patch | POD 4 | Bilateral F, T, P, O | Full recovery |
| Ho et al. 2007 | 33 F | Nothing | PDPH/CS, spinal anesthesia | POD 7 | Bilateral P, O | Full recovery |
| Pugliese et al. 2010 | 41 F | Nothing | Nothing/CS, epidural anesthesia | POD 7 | Bilateral F, O | Full recovery |
| Minai et al. 2011 | N/A F | Nothing | Acute respiratory infection, PDPH/CS, epidural catheter | POD 3 | Bilateral P, O | N/A |
| Orehek et al. 2012 | 26 F | Nothing | Eclampsia, PDPH-like headache, hypertension/CS, epidural anesthesia | POD 5 | Bilateral F, P, O | Mild left arm dysmetria |
| Shah et al. 2014 | 62 F | Hypertension, celiac disease | Hypertension, ileus/laparotomy, epidural anesthesia | POD 4 | Bilateral F, T, O | Minor visual disturbances, memory problems |
| Doherty et al. 2014 | 19 F | Migraine | PDPH/CS, epidural catheter | POD 8 | Lt. F, Rt. T, bilateral P, O | Full recovery |
| Pradhan et al. 2009 | 34 F | N/A | Renal failure, immunosuppression/renal transplant, epidural catheter, blood patch | POD 4 | Bilateral O | Full recovery |
| Hammad et al. 2015 | 72 M | Hypertension | PDPH/laminectomy of L3–S1, spinal drainage, blood patch | POD 15 | Bilateral T, P, O | N/A |
| Shields et al. 2016 | 47 F | Hypertension | Postural headache/T4–T5 discectomy, T2–T8 posterior fusion, spinal drainage | POD 2 | Bilateral F, P, O | Mildly blurred vision |
| Grelat et al. 2014 | 69 F | Subarachnoid hemorrhage | Hydrocephalus/a. depletive lumbar puncture, b. VP shunt | a. 12 hours after surgery, b. immediately | a. Lt. basal ganglia, b. N/A | Hemiplegia, executive function |
| Fok et al. 2016 | 33 F | Obesity | Idiopathic intracranial hypertension/optic nerve sheath fenestration, lumbo-peritoneal shunt | POD 4 | Bilateral P, O | Decreased vision |
| Moriarity et al. 2001 | 19 M | Nothing | Hydrocephalus, VP shunt, BP instability/resection of 4th ventricular tumor | immediately | Bilateral P, O | Mildly conjugated gaze |
| Patel et al. 2010 | 6 M | Nothing | Hydrocephalus, ventricular drain, BP instability/resection of cerebellar tumor | immediately | Bilateral F, T, P, O, thalami, cerebellum, brainstem | Full recovery |

Contd...
all of the above-mentioned reports directly described a relationship between ICP and PRES, these cases indicate a strong association between PRES and a rapid reduction in ICP. In fact, cases with a high risk of severe CSF leak, such as in epidural catheter migration,[23] continuous lumbar drainage after spinal surgery,[24] and lumbar drainage for incisional effusion after spinal surgery,[25] were reported to antedate PRES, which is similar to the situation described here in Case 1. In addition, the emergence of PRES after surgeries, such as VP shunt,[12] lumbo-peritoneal shunt,[26] and posterior fossa tumors with obstructed hydrocephalus,[2,11,23,26,31] have also been reported.

In Case 2, the cysto-peritoneal shunt for the large arachnoid cyst might have caused an acute drop in ICP that resulted in PRES, a novel situation that has not been reported in the literature, to the best of our knowledge. Theoretically, several neurosurgical procedures are at high risk for unintentional hyperperfuasions. Although rare, surgeons should be aware of the possibility of PRES in patients with severe headache, seizure, and disturbances in consciousness that cannot be explained by other medical conditions.

The prognosis of PRES is generally good and almost all patients make full recoveries or return to a normal life with minor deficits.[13] Therefore, we believe that a greater recognition of PRES and careful conservative management, such as avoidance of excessive CSF drainage and early antiepileptic treatment, are critical to minimize complications from this serious but treatable condition.

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Conflicts of interest
There are no conflicts of interest.

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