

Reversible Posterior Leukoencephalopathy Occurring during Resection of a Posterior Fossa Tumor: Case Report and Review of the Literature

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OBJECTIVE AND IMPORTANCE: Our goal was to present a clinically and radiographically documented case of reversible posterior leukoencephalopathy (RPL) that occurred during resection of a posterior fossa tumor. Although RPL has been previously described in multiple nonsurgical settings, we hope that this case description makes RPL more clinically and radiographically recognizable to neurosurgeons.

CLINICAL PRESENTATION: RPL is the clinical syndrome of headaches, altered mental status, seizures, and visual loss, with radiographic findings of reversible parieto-occipital changes on cerebral computed tomographic and magnetic resonance imaging scans. It has been previously reported in the settings of malignant hypertension, renal disease, eclampsia, and immunosuppression. To our knowledge, the patient presented represents the first clinically and radiographically documented case of RPL occurring during resection of a posterior fossa tumor. The patient intraoperatively exhibited wide fluctuations in blood pressure and awoke with clinical and radiographic findings consistent with RPL.

INTERVENTION: Aggressive intraoperative and postoperative management of the patient's blood pressure, supportive intensive care, rehabilitation, and close radiographic follow-up were performed.

CONCLUSION: RPL can occur as a result of intraoperative variations in blood pressure, even among young, previously healthy individuals. With the aforementioned interventions, the patient experienced significant clinical and radiographic recovery. (Neurosurgery 49:1237–1240, 2001)

Key words: Ependymoma, Hypertensive encephalopathy, Occipitoparietal encephalopathy, Posterior fossa surgery, Reversible posterior leukoencephalopathy

In the late 1920s, the term *malignant hypertension* was first applied to patients with hypertension causing papilloedema and retinal hemorrhage (6). Also at that time, Oppenheimer (8) introduced the term *hypertensive encephalopathy* to describe the syndrome of head-

aches, altered mental status, and seizures. In 1996, Hinchey et al. (5) defined reversible posterior leukoencephalopathy (RPL) as a combination of headaches, altered mental status, seizures, and visual loss, with the radiographic findings of predominantly

white-matter changes in the parieto-occipital region. Only 1 of the 15 patients described by Hinchey et al. (5) was younger than 18 years of age. However, RPL was subsequently also observed among pediatric patients (9).

The syndrome of RPL has been observed in multiple clinical settings. Although the majority of these settings involve significant elevations in blood pressure (malignant hypertension, hypertensive encephalopathy, eclampsia, or acute glomerulonephritis), multiple cases have been observed in the absence of hypertension (immunosuppression, interferon therapy, or renal insufficiency).

Radiographically, patients bilaterally exhibit low attenuation on computed tomographic scans and high signal intensity on T2-weighted magnetic resonance imaging (MRI) scans, in the parieto-occipital region (10). The findings are frequently confined to the region of the posterior circulation but often with sparing of the midline portion of the occipital lobes. As the name implies, white-matter findings predominate. However, even in the original description by Hinchey et al. (5), 4 of the 15 patients exhibited radiographic changes involving the gray matter as well (5). The high frequency of accompanying gray-matter changes (1, 9, 15) has prompted some to question the validity of the term *leukoencephalopathy* (9, 11).

The outcomes of RPL are generally favorable. The majority of clinical and radiographic changes resolve in 2 to 4 weeks, especially with prompt control of blood pressure or cessation of the associated immunosuppressant or chemotherapeutic treatment (5, 9). As with the case presented here, however, there are multiple cases in which some neurological or radiographic sequelae persist as a result of frank infarction (1, 9, 11). To our knowledge, the case we present here is the first clinically and radiographically documented case of RPL occurring intraoperatively.

CASE REPORT

A 19-year-old male patient with an unremarkable medical history presented to his primary physician with

complaints of headaches and vomiting for 6 weeks. He was referred to a local neurosurgeon for evaluation of bilateral papilledema and blurring of vision. Computed tomographic scans revealed hydrocephalus and a fourth-ventricular tumor. The patient underwent ventriculo-peritoneal shunt placement (the cerebrospinal fluid cytological analysis yielded negative results). MRI scans obtained after shunting revealed resolution of the hydrocephalus and a heterogeneously enhancing fourth-ventricular tumor (Fig. 1). During the examination, the patient was alert and conversant, with only mild dysmetria and gait ataxia. He was then referred to our institution for tumor resection.

The patient was positioned prone, with rigid cranial fixation, and a routine midline suboccipital exposure was performed. Retraction of the tonsils revealed a large ependymoma occupying the fourth ventricle. Microsurgical resection was performed in the standard manner.

Two hours after the beginning of the procedure, the patient's cerebellum became significantly swollen. The patient then experienced alternating hypo- and

hypertensive episodes, with pressures ranging from 85/50 to 200/130 mm Hg, for approximately 45 minutes. Dissection was intermittently suspended during this period, while the anesthesiologist managed the patient's blood pressure with Neo-Synephrine (Bayer Corp., Morris Township, NJ), esmolol, and labetalol. In addition, the flexion of the patient's neck was reduced, his head was elevated, mannitol was administered, and the patient underwent hyperventilation. After the cerebellar swelling resolved and the blood pressure normalized, a gross total resection was performed without further incident.

While emerging from anesthesia, the patient experienced two generalized tonic-clonic seizures. Emergency computed tomographic scans revealed an open fourth ventricle, no residual tumor bulk, no hematoma, and no hydrocephalus. However, there was bilateral low attenuation in the parieto-occipital regions (Fig. 2). The patient was treated with phenytoin. An electroencephalogram obtained at that time was negative for epileptiform activity. MRI scans performed 4 days later revealed bilateral T2-weighted signal hyperintensity involving the parietal and occipital lobes. Both gray- and white-matter signal changes were noted (Fig. 3).

Postoperatively, the patient was lethargic for 5 to 7 days, with intermittent headaches and patchy visual loss. His seizures were pharmacologically controlled, and he remained normotensive. Ten days after surgery, the patient was transferred to an inpatient rehabilitation facility, where he exhibited steady improvement.

Eight weeks after tumor resection, MRI revealed significant resolution of the

parieto-occipital changes (Fig. 4). However, regions of cortical laminar necrosis, suggesting infarction, were also noted (Fig. 5). On examination, the patient was conversant, ambulatory, and headache-free. He continued to exhibit a mildly disconjugate gaze, but an ophthalmological examination revealed full visual fields. The patient has remained seizure-free while receiving anticonvulsants.

DISCUSSION

To our knowledge, this case represents the first clinical and radiographic description of a posterior fossa tumor. Postoperatively, our patient manifested all of the clinical characteristics of RPL, including headaches, visual loss, seizures, and mental status changes. In addition, scans obtained in the immediate postoperative period demonstrated bilateral parieto-occipital areas of low radiodensity (Fig. 2) and T2-weighted signal hyperintensity (Fig. 3). The mixed involvement of gray and white matter, the anatomic distribution of the changes, and the bilateral symmetry were identical in appearance to other cases of RPL (1, 5, 9, 10). Furthermore, the sparing of the midline occipital region and the lateral extension of the changes (beyond the watershed region) suggest that simple posterior circulation infarction is less likely. Follow-up T2-weighted scans demonstrated significant resolution of the parieto-occipital changes (Fig. 4). However, there were persistent areas of focal encephalomalacia and T2-weighted signal hyperintensity. Corresponding T1-weighted scans demonstrated linear signal hyperintensity consistent with cortical laminar necrosis

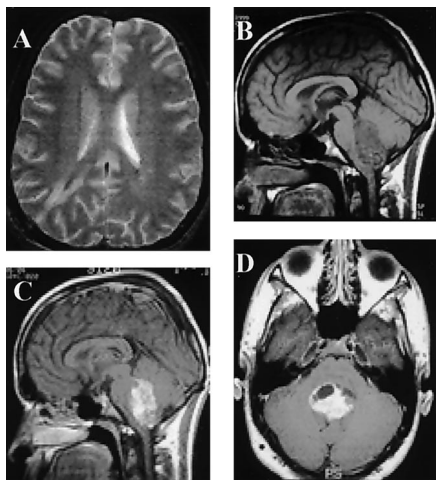


FIGURE 1. MRI scans obtained after shunting. A T2-weighted scan obtained after placement of a right parieto-occipital shunt revealed resolution of the hydrocephalus (A, axial view). T1-weighted scans obtained without (B, sagittal view) and with (C, sagittal, and D, axial views) gadolinium administration demonstrated an enhancing mass filling most of the fourth ventricle.

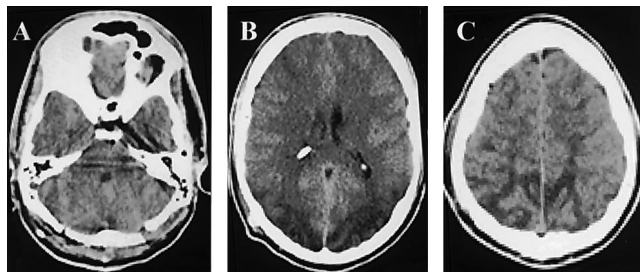


FIGURE 2. Immediately postoperative, axial computed tomographic scans without enhancement. The fourth ventricle appeared to be devoid of tumor or blood clot (A), and

there was no evidence of hydrocephalus (B and C). Regions of low radiodensity were present in both parieto-occipital regions (B and C) and appeared to involve both white and gray matter.

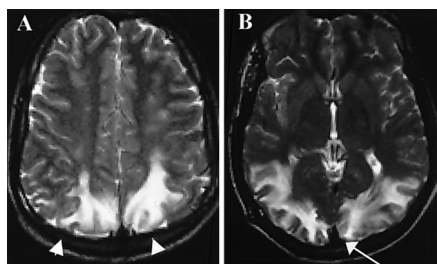


FIGURE 3. Axial, T2-weighted, MRI scans obtained 4 days after tumor resection, revealing bilateral parieto-occipital areas of high signal intensity involving both gray (*arrowheads*) and white matter. There was no significant mass effect, and the superior sagittal sinus remained patent; a normal flow void was demonstrated (*arrow*).

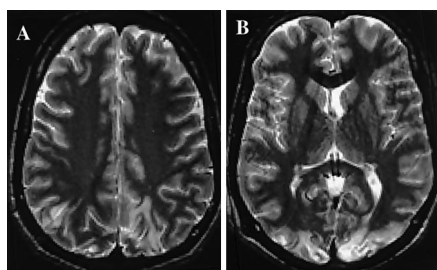


FIGURE 4. Axial, T2-weighted, MRI scans obtained 8 weeks after tumor resection. Bilateral areas of T2-weighted signal hyperintensity involving the parietal and occipital lobes could again be observed. In comparison with the previous examination results (*Fig. 3*), there was significant resolution. The areas of greatest signal resolution were in the white matter.

(*Fig. 5*). This constellation of findings was thought to represent partial resolution of the patient's posterior leukoencephalopathy, with some focal areas of infarction.

The pathophysiological features of RPL remain unknown. For cases in which hypertension is thought to be the inciting event, two main theories predominate. The first suggests that breakthrough vasodilation occurs as the blood pressure exceeds the autoregulatory capability of the cerebral vasculature. This is consistent with the tendency for the edema pattern to be primarily vasogenic (largely sparing gray matter), reversible, and involving

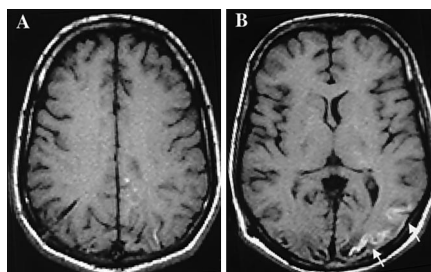


FIGURE 5. Axial, T1-weighted, non-contrast-enhanced, MRI scans obtained 8 weeks after tumor resection, demonstrating linear areas of T1-weighted signal hyperintensity that corresponded to the cerebral cortex and primarily involved the left occipital and parietal regions (*arrows*). There was no significant mass effect.

the posterior vascular distribution, which has less sympathetic innervation (3, 4, 7, 12, 13). A second proposed mechanism involves reflex cerebral vasoconstriction in response to supranormal systemic pressure. This has been demonstrated angiographically and would explain cases in which there is involvement of gray matter and occasional progression to infarction (2, 12, 14). In contrast, non-hypertensive cases (related to immunosuppressant or chemotherapeutic treatment, for example) are possibly caused by alterations in the blood-brain barrier or cytotoxic effects on the cerebrovascular endothelium (5).

CONCLUSION

We describe a case of RPL that occurred during routine resection of a posterior fossa tumor. Although RPL has been previously described in multiple nonsurgical settings, we hope that this case description makes RPL more clinically and radiographically recognizable to neurosurgeons.

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COMMENTS

The authors report a case of a patient with reversible posterior leukoencephalopathy (RPL) that occurred during the resection of a posterior fossa tumor. As they point out, this fairly well-described syndrome includes presenting symptoms of headache, mental status changes, visual disturbances, and sei-

zures in conjunction with characteristic, often completely reversible, radiographic white matter changes in the occipital lobes. Although Moriarity et al. report the first association of RPL with the resection of a posterior fossa tumor, the occurrence of RPL in this instance probably was related not specifically to the type of surgery but rather to the hemodynamic instability that occurred during surgery. Nonetheless, neurosurgeons ought to be aware of this neurological entity, and its occurrence in this case as a complication of an intracranial procedure is worth noting.

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Moriarity et al. present an interesting case of a 19-year-old man with 6 weeks of headache and vomiting who was eventually found to harbor a fourth ventricular ependymoma. During surgery, the patient experienced extreme fluctuations in blood pressure and experienced postoperative seizures, altered mental status, and radiographic abnormalities involving predominately the white matter of the posterior parieto-occipital regions. The authors think that this case represents RPL and present it because it occurred in the setting of a posterior fossa tumor resection. They think that this entity, which has been described in conjunction with a number of medical conditions, may be unfamiliar to neurosurgeons. Although the ac-

ronym RPL probably is relatively uncommon in the neurosurgical literature, I think that its underlying concept is well understood by most neurosurgeons. Certainly, the concept of a hyperperfusion syndrome with "leaky" capillaries that produce extensive white matter edema is well known to neurosurgeons, and the tendency for this white matter edema often to be most prominent in the posterior parieto-occipital regions I myself know well. The prototypical condition in which this presentation occurs is hypertensive encephalopathy, although occasionally even neurosurgeons encounter other examples. Although the vasogenic edema within the white matter is certainly reversible, the underlying condition or conditions that led to the edema often can result in more permanent damage. In the case described here, one wonders whether the episodes of hypotension resulted in infarction within areas of the brain that were edematous and had contained perfusion. Regardless of what this entity is called, the key is to remember that it is potentially reversible with meticulous management of blood pressure and meticulous control of intracranial pressure if intracranial pressure, too, is elevated.

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The authors call attention to a rare but important entity, RPL. It is surpris-

ing that an occurrence of this syndrome after any surgical procedure was not described previously. The first sign of trouble in this case was significant cerebellar swelling. The reason for this event is unclear. If the severe hypertension occurred sometime after the swelling had started, then one might consider other causes, such as temporary obstruction of the shunt, impaired venous drainage, or an unrecognized ventilation problem. Fortunately, the authors took the appropriate measures to treat this problem, and the swelling resolved. The preferential distribution of the lesions in the posterior brain region in RPL is not well understood. Ay et al. (1) reviewed the literature regarding this question. The density of sympathetic innervation in the cerebral vasculature is maximal in the internal carotid and anterior cerebral arterial territories. This innervation decreases posteriorly and is least in the basilar artery and its branches. Ay et al. concluded that sympathetic vasoconstriction may be more effective in protecting the anterior circulation from overperfusion in patients with acute hypertension.

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