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Reversible Posterior Leukoencephalopathy Syndrome After Lateral Skull Base Surgery

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Objective: To describe the clinical course, diagnostic features and management of a case of reversible posterior leukoencephalopathy syndrome after a lateral cranial base removal.
Patient: A 58-year-old male patient with an inconspicuous clinical history presented with a lethargic state without localized neurologic deficit in the postoperative period of a subtotal petrosectomy for an adenoid cystic carcinoma of the temporal bone.
Interventions: Cranial T2-weighted magnetic resonance imaging showed increased signal intensity in the occipital and cerebellar regions, centered at the cortical and subcortical white matter. Antihypertensive drugs, anticonvulsants, and antibiotics were administered. A complete resolution of the symptoms and radiologic findings were achieved within 1 to 2 weeks.
Results: The clinical presentation, radiologic findings, and resolution of the clinical setting are consistent with a diagnosis of reversible posterior leukoencephalopathy syndrome.
Conclusion: To our knowledge, this is the first report of reversible posterior leukoencephalopathy syndrome after a lateral cranial base surgery. **Key Words:** Adenoid cystic carcinoma—Reversible posterior leukoencephalopathy syndrome—Temporal bone tumor.

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Reversible posterior leukoencephalopathy syndrome (RPLS) is characterized by an altered mental status associated with typical neuroimaging findings, showing bilateral high-signal intensity lesions in a T2-weighted magnetic resonance imaging (MRI) (1–3). The etiopathology of RPLS is multifactorial and still uncertain, but it is thought to be related to the failure of cerebral autoregulation leading to the development of vasogenic edema. In most cases, a complete clinical and radiologic recovery occurs. In literature, few cases have been described, with most of them associating RPLS with immunosuppressive treatments (4–6) and elevation of blood pressure (7,8). Cases in relation to intracranial surgery are exceptional.

In this case report, we describe a case of RPLS that occurred in the postoperative period of a subtotal petrosectomy for adenoid cystic carcinoma of the temporal bone. To our knowledge, the case we present here is the first documented case of RPLS occurring after a lateral cranial base surgery.

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CASE REPORT

A 58-year-old male patient was referred to our institution for evaluation and treatment of an adenoid cystic carcinoma of the right external auditory canal with middle ear and intracranial extension. Medical history was unremarkable, and he did not have chronic hypertension. The patient had a 4-year history of complete right facial palsy associated with dysphonia and dysphagia during the last months. The neurologic examination showed right VIIth, IXth, Xth, and XIIth cranial nerve palsy.

The patient underwent a right subtotal petrosectomy. Microsurgical resection included the middle ear, the external auditory canal, the facial nerve, the geniculate ganglion, and the affected middle cranial fossa dura. Occlusion of the right jugular bulb was performed. The surgery and the immediate postoperative period were uneventful. During the fourth postoperative day, the patient showed incisional liquorrhea associated with headache, fever, and dyspnea.

Subsequently, a brain cranial tomographic (CT) scan with uneventful results, a lumbar puncture, and an external lumbar drainage were performed. Cerebrospinal fluid examination revealed no biochemical or microbiologic alterations. Vancomycin and piperacillin-tazobactam were

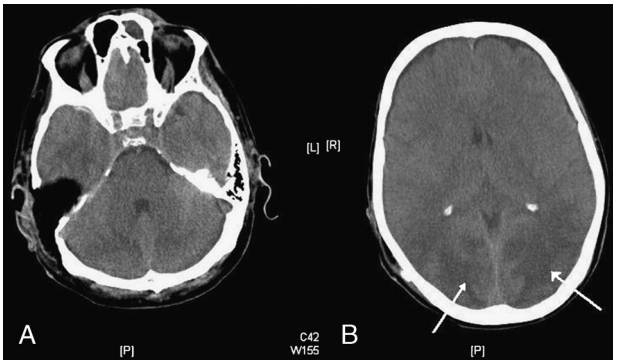

FIG. 1. Axial CT scan shows postpetrosectomy changes with no evidence of hemorrhage (*A*). *Arrows* show areas of hypodensity in the parieto–occipital white matter with mild mass effect (*B*).

started as meningitis prophylaxis. Two days later, the lumbar drainage was removed.

Some hours later, the patient entered a lethargic state. Thorough neurologic examination showed no localized neurologic deficit. Body temperature was 37.5°C, and blood pressure was 170/100 mm Hg. A further brain CT scan (Fig. 1) revealed cerebral edema and bilateral cortical and subcortical hypodensity lesions in the parieto-occipital and cerebellar regions as well as a decrease of the sulcus size; no signs of hemorrhage were found.

The patient was transferred to the intensive care unit where he required orotracheal intubation and mechanic ventilation.

A fluid-attenuated inversion recovery and a T2-weighted MRI were performed on the fifth day. Results showed bilateral high-intensity areas, involving the cortical and subcortical white matter areas with a tendency toward occipital and cerebellar regions, with moderated mass effect, suggesting acute leukoencephalopathy (Fig. 2). Atenolol and nifedipine were used to control hypertension; fenitoin 125 mg per 8 hours was administered as anticonvulsant prophylaxis, and vancomycin, metronidazole, and cefepime were given as antibiotic therapy.

Consciousness was regained, and the mental status recovered gradually within 5 days. The patient was discharged 10 days later, and no sequelae were observed. A 4-month postoperative MRI is shown in Figure 3.

DISCUSSION

RPLS was described by Hinchey et al. (2) in 1996 as a combination of neurologic abnormalities and changes in neuroimaging observed in patients with elevated blood pressure, renal disease, or treatment with immunosuppressive drugs. It also has been described in association with autoimmune diseases, viral or bacterial infections, tumors, trauma, and pregnancy.

Clinical presentation is diverse but usually reflects a global encephalopathy. Headache, disorders of consciousness ranging from drowsiness to stupor, speech or visual disturbance, and seizures are the most common presenting symptoms.

Neuroimaging of RPLS typically reveals bilateral T1-weighted hypointense lesions with a bright cortical and subcortical T2-weighted lesions that affect the white matter and cortex of the parieto–occipital regions (2,9). This signal is not always symmetric and may involve the brainstem, cerebellum, basal ganglia, and frontal lobes. Involvement of the deep gray matter also has been reported (3).

RPLS has been associated with the administration of immunosuppressive and cytotoxic medication, such as cyclosporine, tacrolimus, vincristine, cytarabine, cisplatin (5,10,11), and more recently, bevacizumab (4). These drugs may cause a toxic damage to the vascular endothelium and blood-brain barrier. RPLS also has been related to these drugs because of their hypertensive effects (4).

Several cases have been reported in relation to anesthesia (12), mainly in parturient patients, attributed to the same mechanism as eclampsia and preeclampsia with hypertensive encephalopathy. High blood pressure and disturbance of the blood-brain barrier due to endothelial dysfunction of the cerebral vasculature seem to be the essential causes of RPLS in these patients (13,14).

In the surgical field, Moriarity et al. (15) described a case of RPLS that occurred as a result of blood pressure variation during the resection of a posterior fossa tumor on a healthy patient.

The pathophysiology of RPLS is still uncertain. Hypertensive encephalopathy seems to be the most accepted cause of RPLS. When the blood pressure exceeds the autoregulatory capability of the cerebral vasculature, a disruption of the blood-brain barrier is generated, producing cerebral edema and petechial hemorrhages especially in the posterior regions where there is less density of sympathetic nerves (2,8,12). After a perfusion imaging study in 8 patients, Brubaker et al. (9) suggested that vasogenic edema might be caused by an elevated capillary hydrostatic pressure mediated by venous constriction.

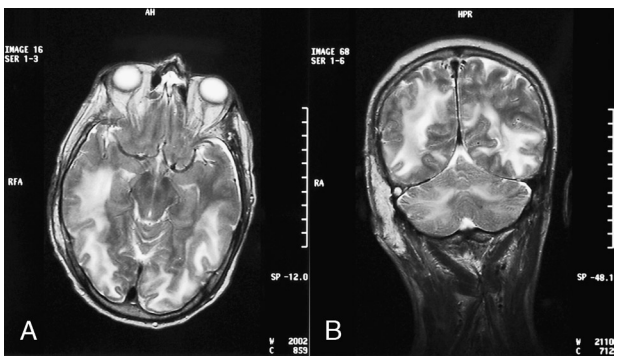

FIG. 2. Cranial magnetic resonance at clinical manifestation time. *A*, Axial T2-weighted magnetic resonance image demonstrates hyperintensity in the white matter bilaterally in the parieto-occipital regions and both cerebellar hemispheres. *B*, Coronal T2-weighted magnetic resonance image shows bilateral areas of T2-weighted signal hyperintensity involving the parietal, occipital, and infratentorial lobes with involvement of the overlying cortex and mass effect.

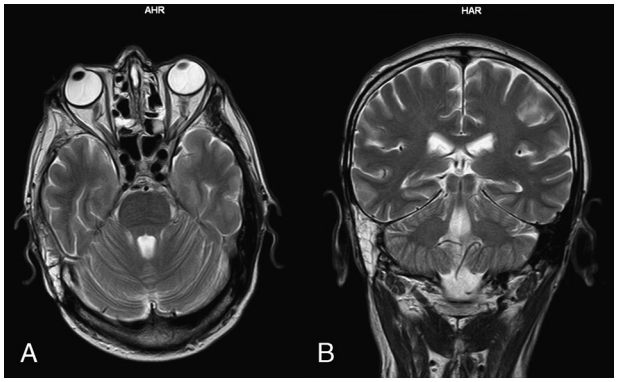

FIG. 3. Axial, T2-weighted, MRI scans obtained after resolution of the syndrome. No significant abnormalities are observed apart from postsurgical changes.

In the present case, no treatment with chemotherapy or immunosuppressive drugs was performed. No electrolyte disturbance was present, and the patient did not have a septicemia. The elevation of blood pressure (170/90) that preceded the first symptoms never reached levels considered to be malignant. However, hypertension cannot be completely ruled out as a cause. RPLS cases (mainly pediatric cases) have been described after minimal changes of the basal blood pressure (8,16).

In our present study, another plausible factor of causing RPLS seemed to be the jugular bulb occlusion, which is performed in many cranial base surgeries. Jugular bulb occlusion is likely to cause severe venous drainage problems especially when the dominant side is affected (17–20). According to the RPLS pathophysiology suggested by Brubaker et al. (9), raised venous pressure also may cause a reduction of intracranial arteriovenous gradient with vasogenic edema and a consequent RPLS. To our knowledge, no reports have so far associated lateral cranial base surgery with the development of RPLS.

Because of its rare incidence, RPLS diagnosis is usually difficult. Neuroimaging findings in the magnetic resonance and clinical symptoms without neurologic focality suggest an encephalopathic syndrome. Favorable outcomes with patient recovery and resolution of the imaging findings are usually achieved and confirm the diagnosis.

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