

Reversible Clinical and Magnetic Resonance Imaging of Central Pontine Myelinolysis Following Surgery for Craniopharyngioma: Serial Magnetic Resonance Imaging Studies

—Case Report—

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Abstract

An 18-year-old girl presented with central pontine myelinolysis (CPM) following surgery for craniopharyngioma. Postoperatively, the patient developed diabetes insipidus with remarkable fluctuation of serum sodium level, suffered a seizure, and developed mental state changes and quadriparesis. Magnetic resonance (MR) imaging obtained soon after the development of the symptoms showed no significant abnormalities. MR imaging obtained 2 months later demonstrated typical trident or bat-like signal abnormalities in the center of the pons, compatible with CPM. Serial MR imaging obtained at 7 and 10 months showed the lesion had decreased in size or almost completely resolved and the patient almost completely recovered. CPM is well known, but neurosurgeons should consider the possibility following surgery for craniopharyngioma.

Key words: central pontine myelinolysis, craniopharyngioma, hyponatremia, prognosis, serial magnetic resonance imaging

Introduction

Central pontine myelinolysis (CPM) is a demyelinating disorder that almost exclusively affects the central portion of the basis pontis and is most commonly found in patients with a history of alcoholism and malnutrition.¹⁻²⁰⁾ Similar symmetrical lesions also occur in extrapontine locations, including the subcortex, thalamus, putamen, and globus pallidus, a condition termed extrapontine myelinolysis.⁹⁾ CPM has been generally observed in patients with a history of alcoholism and malnutrition,¹⁾ but has also been found in patients with anorexia nervosa, orthotopic liver transplantation, diabetes, burns, sepsis, end-stage renal disease, and other diseases.^{3,4,6,7,15,18,20)} Early diagnosis of CPM is often difficult, and some cases are discovered only at autopsy.^{7,13)} The pathogenesis remains unclear, but CPM may be associated with rapid correction or overcorrection of hyponatremia.¹⁹⁾

We report a case of CPM following surgery for craniopharyngioma followed up by serial magnetic resonance (MR) imaging.

Case Report

An 18-year-old girl underwent surgical removal of a craniopharyngioma. Postoperatively, she developed diabetes insipidus (DI), but the urine volume was controlled with desmopressin acetate and serum electrolyte levels were within normal limits. She was discharged after an uneventful postoperative course. However, 6 days following discharge, she was readmitted with vomiting. Initial laboratory values showed hypernatremia (175 mEq/l), and DI had worsened. Although correction of hypernatremia was begun, the serum sodium level fluctuated markedly and normalization was very difficult. Two days after readmission, the patient had a seizure and then developed mental state changes and quadriparesis.

Computed tomography (CT) and MR imaging obtained soon after the development of the symptoms detected no significant abnormalities. Serum sodi-

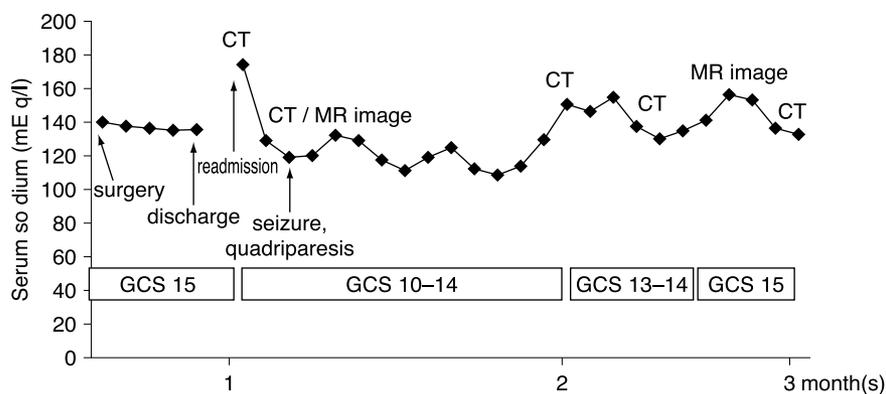


Fig. 1 Postoperative fluctuation of serum sodium levels and clinical course. CT: computed tomography, GCS: Glasgow Coma Scale, MR: magnetic resonance.

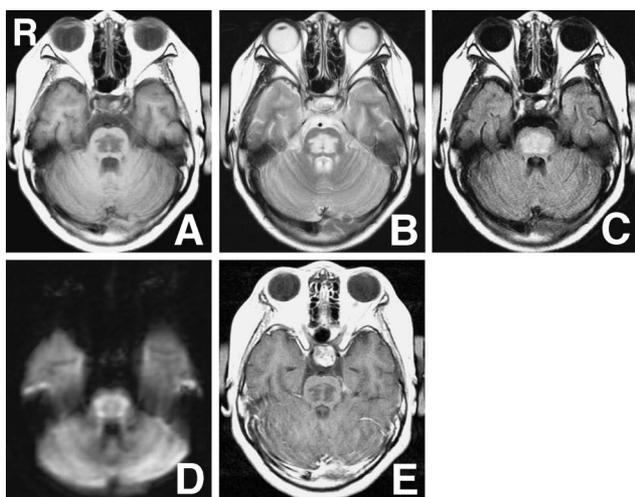


Fig. 2 A: T₁-weighted magnetic resonance (MR) image revealing hypointense abnormality in the central pons. B, C: T₂-weighted and fluid-attenuated inversion recovery MR images revealing typical trident or bat-like hyperintense abnormalities in the same region. D: Diffusion-weighted MR image showing faint hyperintensity. E: T₁-weighted MR image showing the lesion is not enhanced by contrast agent.

um level ranged from 108 to 175 mEq/l during the clinical course. One month later, her symptoms gradually resolved and the serum sodium level normalized (Fig. 1). MR imaging obtained 2 months after the development of the initial symptoms demonstrated symmetrical hypointense abnormalities in the central pons on T₁-weighted imaging (Fig. 2A), and hyperintense abnormalities on T₂-weighted and fluid-attenuated inversion recovery (FLAIR) imaging (Fig. 2B, C). Diffusion-weighted MR imaging

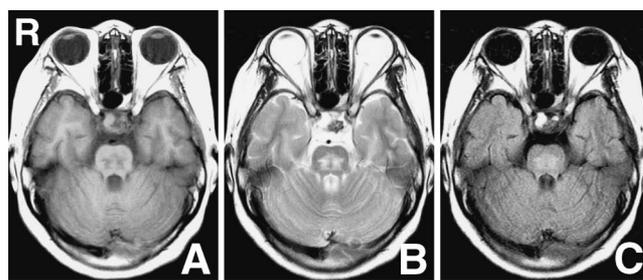


Fig. 3 T₁-weighted (A), T₂-weighted (B), and fluid-attenuated inversion recovery (C) magnetic resonance images obtained 7 months after the initial onset showing the lesion had markedly decreased in size.

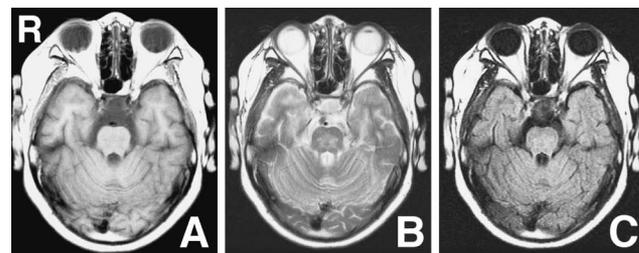


Fig. 4 T₁-weighted (A), T₂-weighted (B), and fluid-attenuated inversion recovery (C) magnetic resonance images obtained 10 months following the initial onset revealing almost complete resolution (A, C) and reduction of the lesion (B).

revealed faint hyperintensity in the same region (Fig. 2D). These findings were compatible with CPM. Serial MR imaging obtained at 7 and 10 months after the initial presentation showed the lesion had decreased in size on T₂-weighted MR imag-

ing and had almost resolved on T₁-weighted and FLAIR imaging (Figs. 3 and 4). The patient had almost completely recovered.

Discussion

CPM may be caused by rapid correction (> 12 mEq/l per 24 hours) or overcorrection of hyponatremia.¹⁹⁾ The sharp increase in serum sodium level results in osmotic endothelial injury that leads to local release of myelin-toxic factors derived from the more vascular gray matter.¹⁴⁾ However, CPM has also been reported in patients with normal or only slightly disturbed sodium level.⁷⁾ Histological studies have shown that oligodendroglial cells are most susceptible to CPM-related osmotic stresses, with the distribution of CPM changes paralleling the distribution of oligodendroglial cells.^{14,16)} Conversely, osmotic demyelination syndrome developing after correction of hyponatremia was complicated by hypokalemia in 89% of patients, which is associated with decreased concentrations of sodium- and potassium-activated adenosine triphosphatase in endothelial or glial cell membranes.¹⁰⁾ Several mechanisms are possible, such as physical shearing of myelin from the axon due to cell shrinkage and/or effects of localized perivascular edema after disruption of the blood-brain barrier,^{9,17)} but how these mechanisms lead to selective myelin loss remains unclear.

In the present case, the tumor was subtotally removed but the calcified component and pituitary stalk were preserved. Although the patient was discharged after an uneventful postoperative course, the patient was readmitted after one month because of hypernatremia caused by hydration due to worsening of DI. Normalization of serum sodium was very difficult despite careful management. We suggest that the fluctuation of sodium levels may have lead to the CPM. The patient's inexperience with self-management using desmopressin acetate may have been the ultimate cause of CPM. Therefore, more careful management of DI may be needed.

CPM manifests as various initial signs and symptoms.⁹⁾ The typical clinical presentation includes mental status changes, seizures, bradypnea, quadripareisis, and pseudobulbar palsies, caused by the destructive lesions in the pontine corticospinal and corticobulbar tracts.⁸⁾ Coma or "locked-in" syndrome can also develop in some patients, usually due to the lesion extending from the base of the pons into the tegmentum.^{11,15)}

MR imaging is more sensitive than CT for the diagnosis of CPM.¹⁵⁾ Symmetric hyperintense abnormalities are typically identified in the basis pontis in the acute stage on T₂-weighted and FLAIR MR imag-

ing, and hypointense abnormalities on T₁-weighted MR imaging.^{7,8)} Early diagnosis of CPM is often difficult and some cases were identified only after autopsy.^{7,13)} Diffusion-weighted MR imaging may be useful in the early diagnosis of CPM and can provide valuable prognostic information about the close relationship between apparent diffusion coefficient abnormalities and clinical status.^{2,3,16)} In particular, diffusion-weighted MR imaging may be a better method to identify early pathophysiologic changes because CPM reflects the osmotic disturbance of water/electrolyte balance.

In the present case, diffusion-weighted MR imaging revealed faint hyperintense abnormalities, suggesting that CPM was in the subacute or chronic stage. It is important to note that MR imaging does not always detect lesions within 2 to 3 weeks after appearance of the initial signs and symptoms.^{8,9)} Delayed imaging is recommended to confirm the diagnosis of CPM because some patients may show no clinical symptoms. Furthermore, the diagnosis of CPM should not be excluded in the absence of abnormalities on MR imaging. Routine MR imaging usually identified no abnormalities within a few months following initial presentation.⁷⁾ The utility of serial MR imaging remains unclear in the subacute and chronic stages of CPM. Reduction or complete resolution of these lesions is known.^{4,7,20)} In the present case, CPM was not detected until 2 months after the initial onset of symptoms, and follow-up MR imaging showed reduction or almost complete resolution of the pontine lesions at 7 and 10 months. T₂-weighted imaging is more sensitive to changes in the lesions than T₁-weighted or FLAIR imaging. Improvement in these lesions may represent resolution of acute edema, remyelination, or decreased astrocytic response.²⁰⁾ Early imaging abnormalities in CPM may reflect edema rather than irreversible demyelination of nerve fibers.⁴⁾ Residual abnormalities may represent areas of permanent damage. However, the severity of lesions on imaging often does not correlate with the clinical severity, course, or prognosis.⁷⁾

The differential MR imaging diagnoses of CPM include infarction, multiple sclerosis, encephalitis, glioma, and changes related to radiation therapy and/or chemotherapy.¹²⁾ Additionally, if the lesions are reversible, posterior reversible encephalopathy syndrome should be considered because such lesions have also been localized in the pons.⁵⁾

CPM was initially regarded as a severe and life-threatening disorder.^{1,17)} However, not all cases fall into this category, and reports of asymptomatic and reversible CPM have appeared, in part due to advances in neuroimaging. We believe that the concept

of CPM, which was originally a purely neuropathological entity, has gradually expanded to a clinicoradiological entity, suggesting that major advances in neuroimaging and treatment have greatly modified the prognosis of CPM, which may be much better than previously thought.

Prevention is the most desirable strategy for CPM. Unfortunately, the present patient had intractable DI following surgery for a craniopharyngioma, which resulted in electrolyte abnormalities despite careful management. Although the incidence of CPM may be low, clinicians must always be alert to the possibility of CPM during management for DI following surgery for craniopharyngioma.

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