Subclinical Central Pontine Myelinolysis after Liver Transplantation


CENTRAL PONTINE MYELINOLYSIS (CPM) can occur after liver transplantation leading to high mortality and serious morbidity. The causes are multifactorial in nature. In this study we report a case of CPM found incidentally on the imaging follow-up of a Wilson's disease patient who had undergone living-related liver transplantation (LRLT).

CASE REPORT

A 17-year-old boy was diagnosed with Wilson's disease, with end-stage liver cirrhosis for 5 years. The major indication for LRLT was repeated esophageal variceal bleeding, which needed sclerotherapy and/or banding (eight times between February 1996 and December 1998). A brain magnetic resonance imaging (MRI) was done (one of the workup tests in transplant candidates with Wilson’s disease) which showed a symmetric hyperintensity signal in the bilateral globus pallidus on T1-weighted image. The globus pallidus MRI result may have been due to portosystemic shunting or the paramagnetic effect of copper or iron deposition. The brainstem at this time was unremarkable (Fig 1A).

LRLT was performed on January 14, 1999. The operation was without complications and the patient recovered uneventfully. However, ascites developed and hypoalbuminemia persisted in the postoperative period despite...
administration of albumin and diuretics. Transient elevation of cyclosporine level was noted (around 400 to 600 ng/mL in early February) and, almost concurrently, hypocholesterolemia was also noted (60 to 80 mg%). On March 5, 1999, a followup brain MRI showed a regressive change of the globus pallidus signal (Fig 1B), but a hyperintensity lesion was found at the median raphe of the basis pontis, which was suspected to be central pontine myelinolysis (CPM). On March 18, 1999, a brainstem auditory evoked potential (BAEP) was done and showed some dysfunction of the left-side brainstem. The MRI lesion was actually an incidental finding and could not be clinically predicted, because, throughout the clinical course, neither pseudobulbar palsy nor quadriparesis were noted; the consciousness was unproblematic and clear, although some adjustment problems had been assessed by psychiatric consultation.

Retrospectively, there was a period of time of mild hyponatremia (124 to 128 mEq/L) since an early postoperative day (January 18), which was finally corrected to a normal level 2 weeks later. On April 24, 1999, follow-up MRI showed marked regression of the pons lesion (Fig 1C).

DISCUSSION

CPM is also known as osmotic myelinolysis, a demyelinating disorder that affects almost exclusively the central portion of basis pontis. The cause of CPM remains unclear, but many studies have implicated changes in serum sodium, specifically the rapid correction of hyponatremia or overcorrection of hypernatremia. A subgroup of patients at risk for CPM includes those undergoing orthotopic liver transplantation (OLT), which may be responsible for 10% of the incidence. A unique point of our case include: (1) the patient was nearly asymptomatic, neurologically related to CPM; and (2) it is probably the first case of suspected CPM after LRLT. Although still inconclusive, transient elevation of cyclosporine and hypocholesterolemia may be one of the predisposing factors.

Perioperative fluctuation of serum sodium has been considered the major factor for developing CPM; it is not certain in the present case whether the change of serum sodium is different from previously reported cases because the degree of hyponatremia was not severe and the correction was neither rapid nor excessive.

Although the pathogenesis of CPM in our case is inconclusive, it was demonstrates that it can be developed without overt signs and symptoms, and that even mild hyponatremia may be a risk factor, not necessarily associated with excessive or rapid correction, particularly under liver dysfunction conditions found in the liver transplant patient, with elevated cyclosporine levels and hypocholesterolemia. It may be more common than previously believed if MRI is more requested follow liver transplantation once the aforementioned conditions appear, such as sodium level changes, obvious hypocholesterolemia, or cyclosporine level elevation, even though there is no typical symptoms.

REFERENCES