Evolution of Pontine and Extrapontine Myelinolysis: Clinical Correlation with Serial CT and MRI Studies

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Central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) usually occur in patients with hyponatremia and rapid correction [1–3]. The clinical manifestations have included a subacute onset of conscious disturbance, quadriparesis and cerebellar signs. In earlier studies, CPM and EPM were diagnosed only at autopsy; recently, the diagnosis has frequently been made by CT or MRI [4–6]. Although these conditions are considered to be related with chn-sicity of hyponatremia or a rapid change of osmolarity, the pathogenesis of CPM and EPM still remains incompletely elucidated. We encountered a patient with both CPM and EPM, who had serial imaging studies before and after ‘rapid’ correction of hyponatremia.

Case Report

A 61-year-old man developed three episodes of syncope after daily treatment with 10 mg enalapril and 2 mg trichlormethiazide for hypertension since Dec. 31, 1993. Neurological examination showed confusion with disorientation as to time and place, amnesia, calculation disturbance and unsteady gait. Brain CT scan on Jan. 12, 1994, showed hypodense lesions in the subcortical white matter of both frontal and parietal areas (fig. 1). No abnormality was noted at the basal ganglia or pons. Biochemical studies revealed marked hyponatremia (102 mEq/l) and hypokalemia (2.1 mEq/l) on day 2 after admission. He then received an intravenous saline infusion (1,920 ml/day, 0.9% NaCl) from day 3, and serum concentrations of sodium increased rapidly to 123, 131 and 139 mEq/l on days 4, 6 and 9, respectively. However, on day 10 his condition deteriorated (quadriparesis, difficulties in walking and holding chopsticks, drooling, swallowing disturbance, slurred speech and lack of emotional control). On evaluation, he was nervous and irritable, disoriented as to the time, and also had calculation difficulties and showed personality changes. There were prominent cerebellar signs with dysmetria, dys-synergia, scanning speech, wide-base gait and generalized hyperreflexia. Brain CT scan on Feb. 2, 1994 showed brainstem swelling with loss of visualization of the quadrigeminal cistern. Brain MRI on day 24 disclosed a huge round lesion at the central pons, which was hypointense on T1-weighted images and hyperintense on T2-weighted (fig. 2a) and proton-weighted images. There were also bilateral symmetric hyperintense lesions in the basal ganglia, thalami and subcortical white matter areas of the frontal and parietal lobes on T2-weighted and proton-weighted images (fig. 2b, c). A diagnosis of CPM and EPM was then established.

After supportive treatment, the patient almost completely recovered 2 months later except for a mild terminal tremor in both hands. A follow-up MRI 10 months later showed a decrease of the lesion in the central pons and disappearance of bright signals in the basal ganglia and thalami on T2-weighted and proton-weighted images.

Discussion

The presented case showed mental abnormality due to forebrain lesions before correction of hyponatremia and then cerebellar and long tract signs due to pontine lesions after correction of hyponatremia. Serial neuroimaging studies also revealed that EPM occurred in the subcortical white matter of both frontal and parietal areas before correction of hyponatremia and CPM developed at the center of the pons after a rapid correction of hyponatremia.
Fig. 1. On day 1 of admission, brain CT showed hypodense lesions in the subcortical white matter of bilateral frontal and parietal lobes (arrowheads).

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Fig. 2. Brain MRI on day 24 revealed a huge, round, bright signal over the center of pons on T2-weighted images (a). Symmetric hyperintense lesions in the basal ganglia, parts of the thalamus (b) and subcortical white matter of both frontal and parietal lobes (arrowheads) (c) were noted on proton-weighted images.

recent years this term has, possibly incorrectly, been ascribed to patients with demyelinating lesions involving the pons, extrapontine or both following hyponatremia. In fact, CPM can only represent some of the complications of hyponatremia. Following hyponatremia, two patterns of brain injuries with different mechanisms were suggested. The first entity is brain damage resulting from brain swelling and increased intracranial pressure, which leads to a decline in cerebral blood flow [7, 8]. Cerebral hypoxia may occur, and the lesions are usually confined to extrapontine areas including the basal ganglia, thalami, cerebral peduncles and subcortical areas. Such brain damage often occurs in patients without treatment. The secondary entity is brain damage after rapid correction of hyponatremia due to osmotic injury to vascular endothelial cells resulting in a release of myelino-toxic factors or vasogenic edema [9]. This type of myelinolysis usually occurs at the center of the pons [10]. According to our observation, EPM is not related with therapy for hyponatremia. We thus suggest that EPM may occur first due to cerebral hypoxia, and CPM may follow after a rapid correction of hyponatremia due to osmotic changes which lead to edema and demyelination within the pons.

References