Case Report

Parkinsonism and recovery in central and extrapontine myelinolysis

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Central pontine myelinolysis is a demyelinating affection of central pons diagnosed on the basis of characteristic MRI finding in an appropriate clinical setting. The condition has been described as universally fatal; however, recent reports of recovery have been documented. We report a case of central pontine and extra pontine myelinolysis, which presented with parkinsonian features apart from bulbar symptoms and made a remarkable recovery. A short review of the literature follows.

Key Words: Hyponatremia, central pontine myelinolysis, magnetic resonance imaging

Central Pontine Myelinolysis (CPM) is a demyelinating disease that usually develops following rapid correction of hyponatremia from any cause. It was originally described in chronic alcoholics.[1] Other reported associations include malnourished status, renal failure, diabetes mellitus, and post-orthotopic liver transplantation.[2],[3] However, it can also occur in healthy persons with hyponatremia caused by gastrointestinal or diuretic therapy.[4],[5] Young children and adults of all ages can be affected.[6] The usual presentation is acute onset pseudobulbar palsy with spastic quadriparesis (‘locked in’ syndrome). The condition was earlier described as fatal but recent reports have shown better outcomes. The prognosis is not influenced by the degree of hyponatremia, the size of lesion on MRI or the severity of neurological deficit at presentation. Patients have been reported to survive if the non-specific secondary complications of transient illness are avoided.

Case Report

A 50-year-old hypertensive, non-alcoholic man, who was on diuretics, developed recurrent vomiting followed by altered sensorium. He was admitted in a local hospital and was diagnosed as a case of hyponatremic encephalopathy (Serum sodium level – 116 mmol/l ). He received intravenous sodium supplementation in unmeasured quantity and showed recovery in sensorium. After 72 hours of recovery, the patient developed generalized tremulousness and difficulty in speaking and swallowing, which progressed to a state of mutism and complete dysphagia within two days. Initial MRI brain was normal. For these complaints, he was referred to the Neurology Department and was admitted here. Neurological examination revealed features of bulbar palsy, extra pyramidal rigidity, rest tremor, preserved tendon reflexes and flexor plantar response. His blood investigations revealed mild neutrophilic pleocytosis, normal creatine phosphokinase level, and high sodium level (154mmol/l). CSF examination was normal. After two weeks, the patient was subjected to a repeat MRI brain which revealed symmetrical T2W hyperintense lesions in the central pons (Figure 1) and signal changes in the basal ganglia and midbrain. In this clinical setting, the radiological picture was suggestive of CPM and extra-pontine myelinolysis (EPM). The patient received supportive treatment and dopamine agonist for extra-pyramidal features. After a few days of treatment, rigidity and hypokinesia improved but bulbar dysfunctions were late to respond. On follow-up after six weeks, the patient had completely recovered.

T2 MR image of brain showing characteristic bilaterally symmetrical hyperintensity in central pons

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**Discussion**

The clinical course of affected patients may be biphasic. Following clinical improvement from hyponatremic encephalopathy, a neurological syndrome caused by myelinolysis typically ensues 2 to 3 days after the correction of hyponatremia. This was the pattern in our case as well. Initial symptoms include mutism, dystonia, lethargy and affective changes. Later, the classical symptoms of spastic quadriaparesis and pseudobulbar palsy develop, reflecting damage to the corticospinal and corticobulbar tracts in the basis pontis. These symptoms may be observed in more than 90% of patients. In our case, the patient had mutism and dysphagia, but had no quadriaparesis or pseudobulbar features.

Extrapontine involvement can present with various movement disorders such as ataxia, dystonia and parkinsonism.[1],[6] The reported patient also had EPM involving basal ganglia and the mid-brain which was responsible for masked facies, extra-pyramidal rigidity and rest tremors.

Pathogenesis of myelinolysis and the reasons for vulnerability and predilection of particular brain regions to develop myelinolysis are not fully understood. Pathologically, myelin is destroyed but neurons and axons are typically spared. Classically, the central pons is involved, hence the original name of CPM. This entity is known to affect other areas, notably the thalami, basal ganglia, and cerebral or cerebellar gray-white junctions. The latter condition is termed EPM.[1] In both CPM and EPM, bilateral and symmetrical involvement is usually the rule.[1],[7],[8]

MRI brain is superior to CT scan for the diagnosis of CPM and EPM. MRI brain shows characteristic T2 hyperintense signals in the central pons and basal ganglia bilaterally.[8] The initial images may be normal. The characteristic lesion on MRI appears up to 2 weeks after clinical presentation and involves the entire pons except for a rim of periphery. Radiological differentials include stroke, multiple sclerosis and brainstem glioma. There is no consistent correlation between the persistence of radiographic abnormalities and that of symptoms in surviving patients.[9]

Treatment of myelinolysis is essentially supportive. Although CPM was once believed to be fatal with a survival rate of only 5-10% beyond 6 months, it is now clear that many patients indeed survive much longer.[4],[6] Myelinolysis itself cannot be specifically treated once it develops. The recovery might be due to increased synaptic plasticity leading to compensation or partly due to reversal of neurotransmitter block at the pontine level after resolution of vasogenic edema caused by osmotic endothelial injury and myelinotoxic factors. A number of treatments have been suggested over the years, for example plasmapheresis, IVIG, steroids, TRH, and reinstatement of hyponatremia but none have undergone clinical trials.

A recent large case series documented that outcome does not depend upon the severity of neurological deficit at presentation or on concomitant internal disease including the degree of hyponatremia.[10] MRI findings did not have any prognostic significance. The survival depends upon the proper management and prevention of non-specific secondary complications of transient illness such as aspiration pneumonia, urinary tract infection, septicemia, deep vein thrombosis and pulmonary embolism.

**Conclusion**

CPM is a distinct condition with unique clinical presentations. Physicians should be aware of this entity while dealing with hyponatremic patients. MR imaging is the radiological modality of choice for the diagnosis. CPM is not a universally fatal disease now and long-term survival with proper supportive management has been recently reported.

**References**


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