Letter to the Editor

Recovery of locked-in syndrome in central pontine myelinolysis

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We read, with great interest, the article “Recovery of locked-in syndrome following liver transplantation with calcineurin inhibitor cessation and supportive treatment” by El Moghazy et al. [1]. The authors present an important clinical observation on a favorable recovery of locked-in syndrome due to central pontine myelinolysis (CPM) in association with liver transplantation, hyponatremia, and use of tacrolimus. By presenting a similar case vignette of CPM, we wish to emphasize the hypothesis that recovery is possible and the outcome is not always dismal.

A 43-year-old woman with severe alcoholism developed delayed ataxia, dysarthria, dysphagia, and tetraplegia after correction of hyponatremia. Clinically, a locked-in syndrome was found whereby communication was possible through intact vertical ocular movements. MRI findings were compatible with CPM (Figure 1A–D). The hyponatremia was treated according to published recommendations including intensive care (Figure 1E) [2]. After a rehabilitation program that lasted 4 months, the patient recovered substantially. She is now able to walk without support, communicate adequately, and swallow.

We wish to present our additional case vignette for two reasons. First, the treatment dilemma of hyponatremia is still unresolved. In the literature, this treatment regimen is described as, “Damned if we do, damned if we don’t” [2]. Despite in our patient, a recommended therapy regimen was performed a connection to the sodium replacement was obvious. Thus, the recommendations between 1985 and 2007 tend to define slow sodium replacement, but do not provide clear instruction on how to do it [2,3]. Clinicians caring for patients with hyponatremia should, therefore, be aware of the risk of CPM while replacing sodium. The challenge, however, of the lesser evil between hyponatremic brain swelling and sodium replacement-associated osmotic demyelination syndrome will remain for the time being.

Second, the severe course of disease with marked recovery is probably more frequent than hitherto thought. El Moghazy et al. patient, as well as ours, was diagnosed with severe locked-in syndrome. While early reports describe the outcome of CPM as poor El Moghazy et al. patient and our patient show a good recovery following a profound rehabilitation program. In a recent study of 24 patients with CPM, such a favorable outcome was seen in 60% [4]. Intensive care and rehabilitation programs should, therefore, be undertaken. Beside locked-in syndrome other clinical manifestations present as mental status changes, quadripareasis, pseudobulbar palsy, cerebellar ataxia, or movement disorders [2]. Recently, even asymptomatic cases of CPM have been reported, emphasizing that the clinical spectrum can vary greatly [5].

As El Moghazy et al. point out, the exact etiology of CPM is unclear [1]. A link between rapid correction of hyponatremia and its occurrence has been hypothesized since 1976 and is still undisputed [2]. However, CPM can occur in a setting of quite different clinical conditions, such as liver and renal transplantation, malnourishment, alcoholism, chronic debilitating illness, hypernatremia, hypokalemia, the syndrome of inappropriate antidiuretic hormone hypersecretion (SIADH), hypoglycemia, and others [2]. Thus, El Moghazy et al. statement that CPM may have a multifactorial cause is plausible according to the
current state of knowledge [1]. In their case, a number of potential CPM-causing conditions had been present, such as liver disease, hyponatremia and use of calcineurin inhibitor tacrolimus [1]. Future studies must address the questions of whether or not a common final path exists and what that path is.

Figure 1. Magnetic resonance image (MRI) in central pontine myelinolysis. (A) The axial diffusion-weighted image shows marked signal intensity in the pons. (B) Axial and (C) sagittal T2-weighted images show the lesion in the corresponding region as (D), the coronal FLAIR-sequence. In (E) the subsequent laboratory testing on sodium is given.

Conflict of Interest

The authors declare there is no conflict of interest.

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