Quiz: 45M with difficulty speaking and weakness

Thomas J Pfiffner, MD
Department of Neurology, University of Minnesota

Case

45 year old male was referred by his outside neurologist to the emergency room with difficulty speaking and weakness. He had been previously to another hospital for confusion following an illness in which he "drank a ton of water to stay hydrated." There, his sodium was reported to be 101 mEq/L. He received treatment to correct his electrolytes and was discharged home the next day. Over the next week, he had increasing difficulty speaking with a soft voice, drooling, and decreased facial expression. His hands seemed weak and have been held in a "crunched" posture with tremor, and his walking developed into a shuffle. He also had emotional lability. On exam he had bilateral facial weakness, poor tongue protrusion, static tremor, and bilateral hand weakness. He also had a shuffling gait. He was found to also have dramatic mood swings. When he was admitted to our hospital, we obtained an MRI of the brain. (Figure 1)

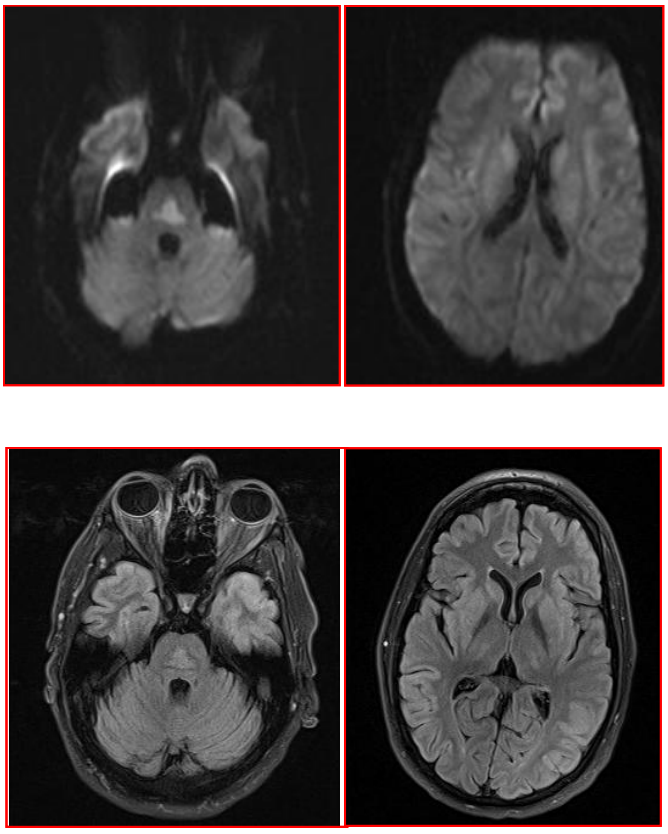

Figure 1: What are the pertinent findings on the MRI diffusion weighted images (DWI; upper row), and Fluid attenuation inversion recovery images (FLAIR; lower row)?

What is your diagnosis?

A. Pontine stroke
B. Central pontine and extra-pontine myelinolysis
C. Basilar artery occlusion
D. Acute disseminating encephalomyelitis
E. Rhombencephalitis

The correct answer is B

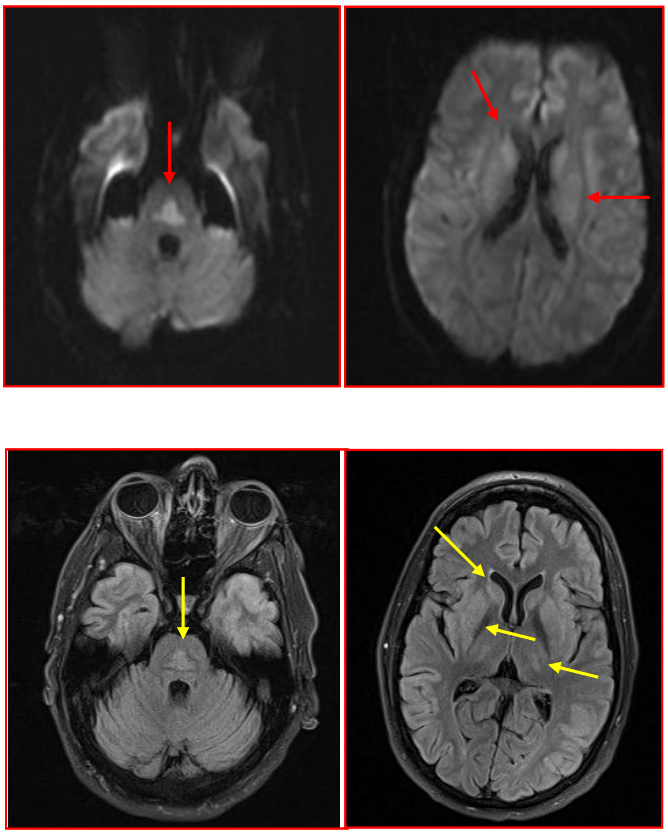

Figure 2: DWI shows diffusion restriction (red arrows), in the central pons and also subtle similar findings in bilateral central thalami, bilateral caudate nuclei and bilateral putamen. There is an associated increased hyperintensity on FLAIR images (yellow arrows) of the same distribution.

Back to the case

The patient's neurological status continued to decline to the point where he could not swallow or talk. Follow up MRI of the brain with and without contrast five days later showed enlargement of the pons lesion and extrapontine lesions (Figure 3).

Patient was treated with supportive therapies. He was discharged 2 weeks after admission with marked dysphagia, hypophonia, bradykinesia, tremor and quadriparesis. Upon return to clinic, approximately 2 months post-discharge, he was found to have made a remarkable recovery. He was able to perform activities of daily living but still had some residual tremor and mood swings.

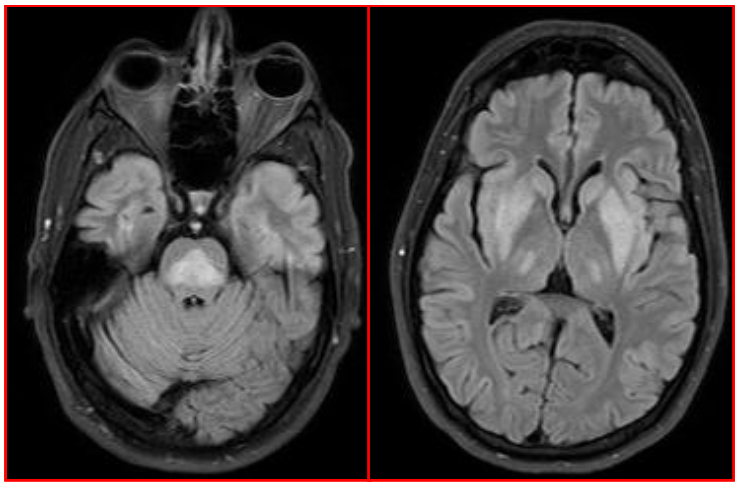

Figure 3: Follow up MRI of the brain with and without contrast 5 days later showed enlargement of the pons lesion and extrapontine lesions

Discussion

The osmotic demyelination syndrome primarily occurs with overly rapid correction of severe hyponatremia that has been present for more than two days. It typically has a biphasic clinical course: 1) encephalopathy from hyponatremia 2) central pontine myelinolysis (CPM)/extrapontine myelinolysis (EPM); dysarthria, dysphagia, paraparesis or quadraparesis, behavior disturbances, lethargy, confusion, disorientation, obtundation, and coma). The clinical manifestations of osmotic demyelination are typically delayed for two to six days after the elevation in the serum sodium concentration. EPM occurs in approximately 10% to 40% of pts with CPM however Parkinsonism is rarely seen in CPM and EPM.

The demyelinating lesions can be detected by CT scanning or, preferably, MRI. However, these tests may not become positive for as long as four weeks after disease onset. Thus, an initially negative radiologic study does not exclude osmotic demyelination. However, earlier detection is possible with newer imaging techniques such as diffusion weighted imaging (DWI).

Treatment is mainly supportive however, immunomodulation (steroids, immunoglobulins, and plasma pharesis) as well as thyrotrophin releasing hormone have all tried and reported as case reports but no trials have been done thus far. Prognosis once thought as poor with mortality 40%-50% now has been reduced markedly with modern neuroimaging allowing for earlier diagnosis . In a most recent series of 34 cases by Menger et al., only 2 died, 1/3 recovered completely, 1/3 was independent with mild residual deficit, and 1/3 was significantly dependent. Prognosis is not influenced by the degree of hyponatremia, size of lesion, or severity of neurological deficit at presentation.

The radiologic differential diagnosis for CPM include infarction, metastasis, glioma, multiple sclerosis, encephalitis, and radiation or chemotherapy. Sparing of the pons periphery favors CPM.Concomitant involvement of the basal ganglia is fairly specific for osmotic myelinolysis. In such cases the imaging differential diagnosis is much more narrow and includes hypoxia, Leigh

disease, and Wilson's disease. Osmotic myelinolysis spares the globus pallidus unlike the others. The classical history combined with imaging findings usually distinguishes osmotic myelinolysis from the rest of the conditions.

References

1. Sajith J, Ditchfield A, Katifi HA. Extrapontine Myelinolysis Presenting as Acute Parkinonism. BMC Neurology 2006; 6:33.
2. Bhoi KK, Pandit A, Guha G, Barma P, Misra AK, Garai PK, Das SK. Reversible Parkinsonism in Central Pontine and Extrapontine Myelinolysis: A Report of Five Cases From India and Review of the Literature. Neurology Asia 2007; 12: 101-109.
3. Sterns, RH, Riggs, JE, Schochet SS, Jr. Osmotic demyelination syndrome following correction of hyponatremia. N Engl J Med 1986; 314:1535.
4. Brunner, JE. Redmond, JM. Haggar, AM, et al. Central pontine myelinolysis and pontine lesions after rapid correction of hyponatremia: a prospective magnetic resonance imaging study. Ann Neurol 1990; 27:61.
5. Ruzek, KA, Campeau, NG, Miller, GM. Early diagnosis of central pontine myelinolysis with diffusion-weighted imaging. AJNR Am J Neuroradiol 2004; 25:210.
6. Osborn, Ann G. Diagnostic Neuroradiology: A Text/Atlas. Elsevier Health Sciences. ed 1, p 763, 1994
7. Menger H, JÖrg J. Outcome of central pontine and extrapontine myelinolysis. J Neurol 1999; 246: 700-5.

Corresponding Author:

Thomas J Pfiffner, MD
Department of Neurology, University of Minnesota
420 Delaware St. SE, MMC 295
Minneapolis, MN 55455
Tel (612) 625-9110, Fax (612) 625-7950
Email: pfiff012@umn.edu