Images in Neurology

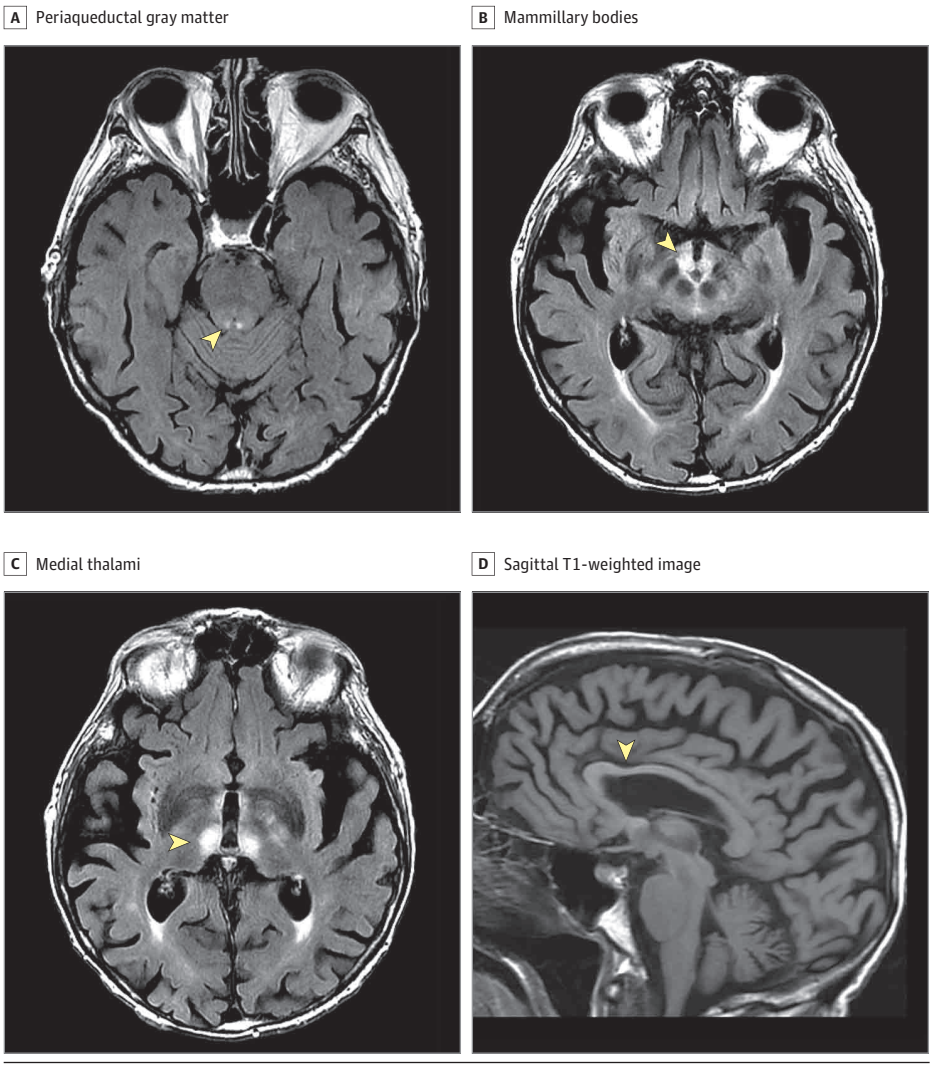
Characteristic Neuroimaging Abnormalities of Korsakoff Syndrome

J. Bradley Segal, BS, BA; Marc A. Bouffard, MD; Gottfried Schlaug, MD, PhD

A man in his 70s with a history of chronically elevated lactate levels, alcoholic cirrhosis, and chronic cognitive decline attributed to hepatic encephalopathy despite lack of asterixis or response to lactulose presented with 1 week of confusion worsened from baseline. On examination, he had anterograde worse than retrograde memory deficits without asterixis, nystagmus, ophthalmoplegia, or ataxia. Findings on magnetic resonance imaging of the brain (**Figure**) were consistent with previously undiagnosed Korsakoff syndrome.[1] His subacute decline was attributed to a urinary tract infection, and the patient's orientation returned to baseline with antibiotics. His lactate level returned to within a normal range with parenteral thiamine, although his memory deficits persisted.

Korsakoff syndrome is an acquired chronic disease characterized by dense anterograde amnesia and short-term memory loss in the setting of otherwise intact cognition.[1] Korsakoff syndrome results from untreated or undertreated Wernicke encephalopathy, an acute, reversible condition caused by thiamine deficiency. Thiamine mediates pyruvate metabolism; its deficiency can result in elevated lactate.[2] Wernicke encephalopathy is most closely

Figure. Magnetic Resonance Imaging Findings of Korsakoff Syndrome



Magnetic resonance imaging shows bilateral fluid-attenuated inversion recovery hyperintensity (arrowheads) in the periaqueductal gray matter (A), mammillary bodies (B), and medial thalami (C). D, Sagittal T1-weighted image demonstrates volume loss in the corpus callosum (arrowhead) and in the cortex with widening of the cerebral sulci.

associated with chronic alcohol abuse but can also arise from nonalcoholic causes of nutritional deficiency such as gastrointestinal surgery.[1]

Only 16.5% of patients with Wernicke encephalopathy present with the classic triad of ataxia, ophthalmoplegia, and confusion.[3] Diagnosis is also frequently confounded by concurrent explanations for a patient's cognitive or motor symptoms such as hepatic encephalopathy or intoxication. As a result, the diagnosis of Wernicke encephalopathy is often missed.[3]

Magnetic resonance imaging is the most valuable study to detect symptomatic thiamine deficiency.[1] Fluid-attenuated inversion recovery sequences demonstrate characteristic symmetric hyperintensities most frequently in the medial thalamus, periventricular region, periaqueductal gray matter, mammillary bodies, and midbrain tectum most likely reflecting edema, loss of neurons, and reactive gliosis.[1] These lesions can be accompanied by symmetrically distributed hyperintensities in atypical locations.[4] When the disease is chronic, T1-weighted imaging can show volume loss in the mammillary bodies and corpus callosum along with enlargement of the sulci and ventricles suggesting atrophy and volume loss.[4]

Once a patient develops Korsakoff syndrome, the prognosis for recovery is poor. Dense amnesia and short-term memory loss are typically irreversible.[1] Disability from Korsakoff syndrome is best prevented by retaining a high index of suspicion when evaluating malnourished patients at risk for thiamine deficiency and immediately administrating parenteral thiamine before glucose if Wernicke encephalopathy is suspected.[1,3]

ARTICLE INFORMATION

Author Affiliations: Harvard Medical School, Boston, Massachusetts (Segal, Bouffard, Schlaug); Department of Neurology, Beth Israel Deaconess Medical Center, Boston, Massachusetts (Bouffard, Schlaug).

Corresponding Author: Marc A. Bouffard, MD, Harvard Medical School, Department of Neurology, Beth Israel Deaconess Medical Center, 330 Brookline Ave, Apt 1, Boston, MA 02118 (marc.a.bouffard@gmail.com).

Published Online: August 29, 2016. doi:10.1001/jamaneurol.2016.1843.

Conflict of Interest Disclosures: None reported.

REFERENCES

1. Sechi G, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. *Lancet Neurol*. 2007;6(5):442-455.

2. Butterworth RF. Thiamine deficiency-related brain dysfunction in chronic liver failure. *Metab Brain Dis*. 2009;24(1):189-196.

3. Harper CG, Giles M, Finlay-Jones R. Clinical signs in the Wernicke-Korsakoff complex: a retrospective analysis of 131 cases diagnosed at necropsy. *J Neurol Neurosurg Psychiatry*. 1986;49(4):341-345.

4. Jung YC, Chanraud S, Sullivan EV. Neuroimaging of Wernicke's encephalopathy and Korsakoff's syndrome. *Neuropsychol Rev*. 2012;22(2):170-180.