Neuroimaging Highlight

Editors: William Hu, Richard Farb

Wernicke Encephalopathy in a Patient with T-Cell Leukemia and Severe Malnutrition

Submitted by: Lynn Lacasse, Cheemun Lum

Can. J. Neurol. Sci. 2004; 31: 97-98

The patient was a 20-year-old male with an eight-month history of T-cell acute lymphoblastic leukemia (ALL). He presented to the emergency room with confusion and decreased level of consciousness that had worsened over the prior two weeks. He complained of generalized fatigue, weakness, intermittent nausea and vomiting and depression. His diet had consisted of only soda pop over the six weeks preceding admission.

The patient was disoriented and had a depressed level of consciousness. He appeared pale, cachectic, and had poor hygiene. Examination of the central nervous system showed lateral nystagmus and slowed motor activity in the both upper and lower extremities. Reflexes were bilaterally 2+ except at the ankles where they were 1+. Plantar responses were bilaterally downgoing.

He had a low Hb (102 g/L) with a mean corpuscular volume of 85.5 fl (80-100), decreased platelets ($103x10^9$/L), lymphocytes ($6.1x10^9$/L) with slight hyponatremia (132 mmol/L) and hypokalemia (3.3 mmol/L). Liver enzymes were elevated (AST 92 U/L, ALT 114 U/L). Drug screen was negative for tricyclic antidepressants, benzodiazepine, ASA, acetaminophen and ethanol.

A computed tomography scan without contrast was normal. Magnetic resonance imaging (MRI) was ordered to exclude encephalitis. An MRI with gadolinium enhancement revealed abnormal high signal on FLAIR images in the periaqueductal region extending superiorly into the dorsal medial thalami and around the third ventricle (Figures 1-2). Abnormal enhancement was seen in the mammillary bodies bilaterally (Figure 3). The MRI suggested an unsuspected diagnosis of Wernicke encephalopathy. The patient was given thiamine 100 mg IV daily for two weeks and progressively improved. The history, imaging findings and response to thiamine strongly suggest the diagnosis

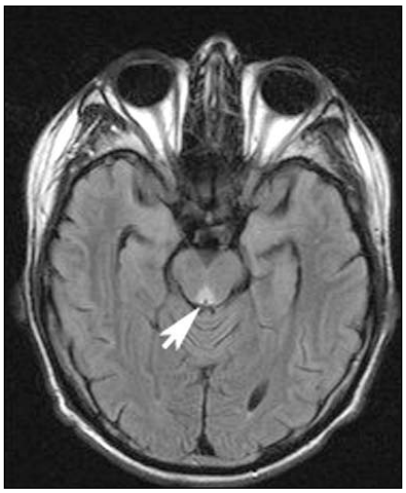

Figure 1: *Axial FLAIR image at level mibrain level demonstrating abnormal high signal in the periaqueductal grey area (arrow).*

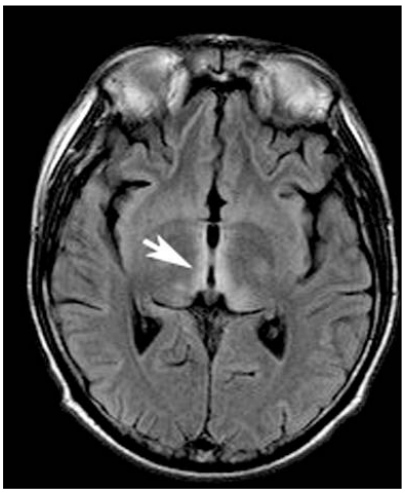

Figure 2: *Axial FLAIR image demonstrates abnormal high signal about the walls of the third ventricle (arrow) and in the thalami.*

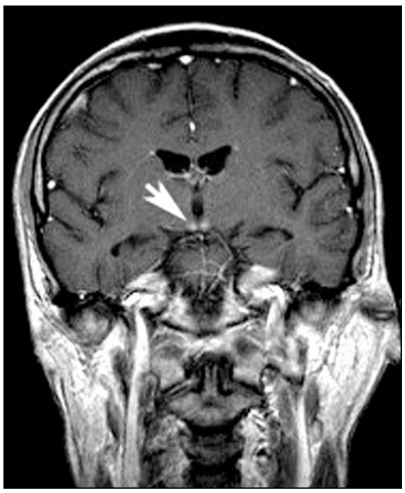

Figure 3: *Coronal T1post gadolinium images demonstrate abnormal enhancement of mammillary bodies (arrow).*

From the Neuroradiology, Division of Radiology, The Ottawa Hospital-Civic Campus, Ottawa, Ontario, Canada.

Received April 23, 2003. Accepted in final form November 17, 2003.

Reprint requests to: Cheemun Lum, Neuroradiology, Division of Radiology, The Ottawa Hospital-Civic Campus, 1053 Carling Avenue, Ottawa, Ontario, Canada, K1Y 4E9.

of Wernicke encephalopathy (WE).

Wernicke encephalopathy is a potentially fatal neurologic disorder caused by thiamine deficiency. If affects the midbrain, mammillary bodies and hypothalamus.[1] Approximately 25-30 mg of thiamine are stored in the body with large amounts found in skeletal muscle, heart, liver, kidneys, and brain.[2] Thiamine may also have a specific role in neurons independent of its function in general metabolism. Requirements are increased in pregnancy, lactation, thyrotoxicosis, and fever. Accelerated losses occur with diuretic therapy, hemodialysis, peritoneal dialysis and diarrhea. Defective absorption occurs in general malabsorption states such as prolonged parenteral nutrition without addition of thiamine and gastrectomy, gastrojejunostomy, alcoholism, chronic malnutrition, and folate deficiency.[3]

The typical MRI findings are symmetrical high-signal lesions surrounding the third ventricle and aqueduct on T2 or proton density-weighted images. Pathologically, the acute stage is characterized by marked vascular dilatation, endothelial swelling, and neuronal damage.[4] Classical MRI findings in WE demonstrate diffuse enhancement of the mammillary bodies, the quadrigeminal plate, the periaqueductal grey matter, and the thalami.[3,5,6] Recent studies have demonstrated cases of acute WE demonstrating abnormalities at diffusion-weighted imaging.[7-9] Restricted diffusion may represent neuronal necrosis and may suggest a poorer response to therapy than patients demonstrating normal or increased diffusion.[8] Unfortunately, diffusion-weighted imaging was not performed on this patient and attempts to recall the patient for this sequence and for follow-up imaging were unsuccessful.

Chronic WE may demonstrate atrophy on MRI consistent with reabsorbed and reorganized petechial hemorrhage of the affected areas.[4] In chronic lesions there is loss of neuropil with fibrillary astrocytosis and atrophy of the region involved.[10]

One explanation for the typical imaging and pathological findings in acute and chronic WE suggests that the periventricular regions have a higher rate of thiamine-related glucose and oxidative metabolism. This renders these areas more susceptible to changes in thiamine-deficient states.[11]

Wernicke encephalopathy is a potentially fatal metabolic disorder which is fully reversible with thiamine. However, early diagnosis is essential to prevent irreversible neuronal damage. In our case, severe malnutrition and personal neglect contributed to thiamine deficiency. An MRI may show characteristic findings and suggest the diagnosis in previously unsuspected cases.[6]

References

1. Gallucci M, Bozzao A, Splendiani A, et al. Wernicke encephalopathy: MR findings in five patients. AJNR 1990; 11:887-892.
2. Wilson JD. Vitamin deficiency and excess. In: Fauci AS, Braunwald E, Isselbacher KJ, et al (Eds). Harrison's Principles of Internal Medicine, 14th Edition: McGraw-Hill 1998: 482-483.
3. D'Aprile P, Tarantino A, Santoro N, Carella A. Wernicke's encephalopathy induced by total parenteral nutrition in patient with acute leukemia: unusual involvement of caudate nuclei and cerebral cortex on MRI. Pediatric Neuroradiology 2000; 42:781-783.
4. Park SH, Manho K, Duk LN, Beom SJ. Magnetic resonance reflects the pathological evolution of Wernicke encephalopathy. J Neuroimaging 2001; 11:406-411.
5. Shogry MEC, Curnes JT. Mamillary body enhancement on MR as the only sign of acute Wernicke encephalopathy. AJNR 1994; 15:172-174.
6. Antunez E, Estruch R, Cardenal C, et al. Usefulness of CT and MR imaging in the diagnosis of acute Wernicke's encephalopathy. AJR 1998; 171:1131-1137.
7. Doherty MJ, Watson NF, Uchino K, Hallam DK, Cramer SC. Diffusion abnormalities in patients with Wernicke encephalopathy. Neurology 2002;58:655-657.
8. Bergui M, Bradac GB, Zhong J. Diffusion abnormalities and Wernicke encephalopathy. Neurology 2003;60:727-728.
9. Bergui M, Bradac GB, Zhong JJ, et al. Diffusion-weighted MR in reversible Wernicke encephalopathy. Neuroradiology 2001;43:969-972.
10. Brody BA.The Wernicke-Korsakoff syndrome: neuropathology and pathogenic basis. Int J Neuroradiology 1996; 2:216-230.
11. Witt ED. Neuroanatomical consequences of thiamine deficiency: a comparative analysis. Alcohol Alcohol 1985;2:201-221.

https://doi.org/10.1017/S0317167100002894 Published online by Cambridge University Press