Wernicke's Encephalopathy: Report of a Case

Verajit Chotmongkol, MD*,
Panita Limpawattana, MD*

** Department of Medicine, Faculty of Medicine, Khon Kaen University, Khon Kaen*

A case of Wernicke's encephalopathy is reported. A 53-year-old alcoholic man was presented with acute confusion, upward gaze palsy of the left eye and gait ataxia for 5 days. The typical findings upon MRI of the brain showed multiple, scattered, small non-enhanced low signal intensity on T1WI, high signal intensity on T2WI involving at bilateral basal ganglias, and thalami, midbrain, pons, and periventricular regions with evidence of atrophic changes of brain and the mamillary bodies. After treatment with intravenous thiamine 100 mg/day, the left upward gaze palsy was fully recovered at the 8th day of treatment. The mental status and gait ataxia gradually improved.

Keywords: *Wernicke's encephalopathy*

J Med Assoc Thai 2005; 88(6): 855-8
Full text. e-Journal: *http://www.medassocthai.org/journal*

Wernicke's encephalopathy (WE) is a serious neurological disorder that is uncommon, and can be fatal if not treated. The incidence varies due to misdiagnosis. Certain studies showed a prevalence of 0.8% in normal population and 12.5% in alcoholism[1,2]. The classic clinical triad includes acute onset of encephalopathy, ataxia and ophthalmoplegia, which is found in only one-third of the patients upon initial examination[3]. The mortality rate is 10-20%; therefore prompt recognition and treatment are very important. In the present paper, the authors reported a case of WE with typical presentation.

Case Report

A 53-year-old man was admitted to Srinagarind Hospital in September 2003 with the chief complaint of acute confusion and gait ataxia for 5 days. He had been an alcoholic for 33 years and was still drinking alcohol at the time of this illness. He had a motorcycle accident 10 years ago with residual impairment of eye vision, and ptosis of the left eye and right facial nerve palsy.

Physical examination showed a drowsy and disorientated man with a body temperature of 38.0°C. Neurological examination revealed ptosis, an oval-shaped, and unreactive to light pupil and upward gaze palsy of the left eye, right facial palsy (LMN type), and gait ataxia. He also had positive signs of chronic liver disease. Other findings were unremarkable.

The complete blood count, serum glucose, BUN, creatinine, electrolytes, urinary analysis and chest X-ray were within normal limits. Liver function test revealed an aspartase aminotransferase level of 90 U/L. The MRI of the brain showed multiple, scattered, small non-enhanced low signal intensity on T1WI, high signal intensity on T2WI involving at bilateral basal ganglias, thalami, midbrain, pons, and periventricular regions (Fig. 1) with evidence of brain atrophy. The mamillary bodies could not be identified. A cerebrospinal fluid analysis was within normal limits. The electroencephalograph (EEG) showed background alpha and theta intermixing activities without epileptic discharge.

WE was diagnosed. The patient was treated with intravenous thiamine 100 mg once daily for WE and 1.2 mg of amoxi-clavulonic acid every 8 hours for aspiration pneumonia. The results of hemocultures, sputum culture and CSF culture were negative. His body temperature returned to normal at the 3rd day of treatment. The left upward gaze was fully recovered at the 8th day of treatment. The mental status and gait ataxia gradually improved. He was referred to a local hospital and lost to follow-up.

Correspondence to: Chotmongkol V,Department of Medicine, Faculty of Medicine, Khon Kaen University, Khon Kaen 40002, Thailand.

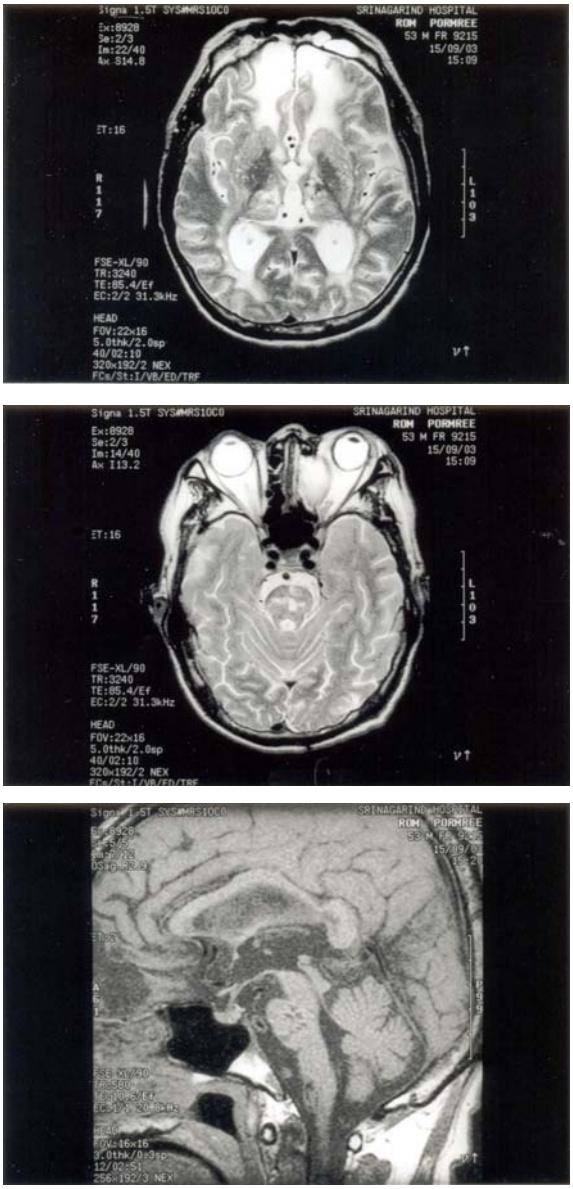

Fig. 1 MRI of the brain revealed multiple, scattered, small high signal intensity on T2WI involving at bilateral basal ganglias, thalami, midbrain, pons, and periventricular regions with evidence of brain atrophy. The mamillary bodies could not identify on T1WI

Discussion

WE is a medical emergency caused by thiamine or B1 deficiency. The most common cause is found in alcoholism but it is also common in prolonged intravenous feeding, fasting, refeeding after starvation, prolonged vomiting such as hyperemesis gravidarum, anorexia nervosa, patients receiving dialysis, thyrotoxicosis, congestive heart failure with long term diuretic treatment, and AIDS[1-8]. Glucose administration can precipitate WE in patients who have thiamine deficiency. However, it is also caused by a genetic abnormality resulting from a defection in transketolase structure that diminished binding with coenzyme thiamine pyrophosphate[3]. Thiamine (B1) is a cofactor for transketolase, alpha-ketoglutarate dehydrogenase, pyruvate dehydrogenase, and branch-chain alpha-keto acid dehydrogenase that are essential in intermediate carbohydrate metabolism. Thiamine may also function in axonal conduction and synaptic transmission. Thiamine deficiency causes alteration of cerebral energy mechanisms, diminishes nerve impulse transmission at synapses and impairs DNA synthesis that characterized by necrosis of nerve cells and myelinated structures. WE may evolve as minor episodes comprising combinations of these more subtle features and that the clinical form of the disease develops after repeated episodes of subclinical encephalopathy, which is supported by autopsy findings. The characteristic brain lesions include symmetrical discoloration of structures surrounding the third ventricle, aqueduct, and fourth ventricle, with petechial hemorrhages in occasional acute cases and atrophy of the mamillary bodies in a majority of chronic cases. Involvement of the third and sixth nuclei and tegmentum account for the ophthalmoplegias. The average age at onset among adults is 50 years and male-to-female ratio is 1.7:1 and 3-4:1 in alcoholism[3]. The classical triad is characterized by ocular abnormalities, ataxia and a global confusion state that are found in only one - third of patients upon initial examination. It must be reemphasized that the onset of the disease may be acute, subacute, or chronic. The global confusional state may include apathy, impaired awareness of the immediate situation, spatial disorientation, inattention and inability to concentrate. Coma is a particularly important feature for early recognition of WE, since it may be the sole manifestation. The common ocular abnormalities are horizontal nystagmus and lateral rectus muscle palsies. The other findings are conjugate-gaze palsies, ptosis, pupillary abnormalities, complete ophthalmoplegia, and papilledema. It can be one-sided, or on both sides. The loss of equilibrium seen in the early stages is due to vestibular paresis while the wide-based ataxic gait seen in the subacute and chronic phases is due to cerebellar dysfunction[1].

Diagnosis of WE has been based on history, clinical symptoms and the laboratory investigations such as low serum thiamine and transketolase activity

and elevation of pyruvate level[4,5]. The CT scan of the brain occasionally shows low attenuation with or without enhancement in the involved regions, especially with symmetric low density in the thalami[2,7]. The MRI of the brain has now considerably been used as a diagnostic adjunct in the patient suspected of WE[7,9-11]. The typical MRI finding in acute phase is low signal intensity on T1-weighted images and increased signal intensity on T2-weighted images located in the bilateral thalami, periaqueductal areas of the third ventricle, floor of the fourth ventricle, tectal area of the midbrain, mamillary bodies, and cerebellar vermis. Certain patients show subtle enhancement from postcontrast T1-weighted images. Mamillary enhancement alone has been reported as the only acute sign[7]. In the late phase, T2 hyperintensity disappears and focal volume loss with gliosis results in mamillary atrophy, and dilatation of the third ventricle and aqueduct[10].

The mortality rate is 10-20% and the complications are permanent loss of memory and cognitive skills. The treatment may correct all abnormalities so it must be viewed as a medical emergency. The goal of treatment is to control symptoms as much as possible and to prevent progression of the disorder. Thiamine is the drug of choice for treatment of WE. Dosage of 2 mg intravenous may be enough to reverse the ocular symptoms which generally begin to improve within hours to days, but initially at least 100 mg is recommended. Ataxia and confusion may also resolve dramatically, although improvement may not be noted for days to months. Glucose administration must be given after thiamine administration and continued until the patients can eat a normal diet. Total abstinence from alcohol is required to prevent progressive loss of brain function.

In the presented patient, although neither the serum thiamine nor transketolase activity were available, WE was most likely recognized due to a history of chronic alcohol consumption with typical abnormal neurological signs and MRI findings in acute and chronic phase of WE as mentioned above, and finally, he had a dramatic response to thiamine treatment. From this presentation, WE should be looked for in this clinical setting.

References

1. Charness ME, Simon RP, Greenberg DA. Ethanol and the nervous system. N Engl J Med 1989; 321: 442-54.
2. Hemphill JC, Beat MF, Gress DR. Critical care neurology. In: Braunwald E, Fauci AS, Kasper DL, Hauser SL, Longo DL, Jameson JL, eds. Harrison's principle of internal medicine, 15th edition. New York: McGraw-Hill, 2001: 2491-5.
3. Philip Salen, Emedicine - Wernicke's encephalopathy. http://www.emed.com/emerg/topic642.htm.
4. Medlineplus Med Encyclopedia: Wernicke - Korsakoff Syndrome. http://www.nln.nih.gov/medlineplus/encyclopedia/article/000771.htm.
5. Wernicke - Korsakoff Syndrome. http://www.rwjhamilton.org/Atoz/encyclopedia/article/000771.asp.
6. Kinsella LJ, Riley DE. Nutritional deficiencies and syndromes associated with alcoholism. In: Goetz CG, ed. Textbook of clinical neurology, 2nd edition. Philadelphia: Saunders, 2003: 873-93.
7. Ball WS Jr, Degrauwn A. Metabolic, congenital neurodegenerative, and toxic disorders. In: Edelman RR, Hesselint JR, Zlatkin MB, eds. Clinical magnetic resonance imaging, 2nd edition, Philadelphia: WB Saunders, 1996: 880-910.
8. Hemphill JC, Beat MF, Gress DR. Critical care neurology. In: Braunwald E, Fauci AS, Kasper DL, Hauser SL, Longo DL, Jameson JL, eds. Harrison's principle of internal medicine, 15th edition. New York: McGraw-Hill, 2001: 2491-5.
9. Chung SP, Kim SW, Yoo IS, Lim YS, Lee G. Magnetic resonance imaging as a diagnostic adjunct to Wernicke encephalopathy in the ED. Am J Emerg Med 2003; 21: 497-502.
10. Park SH, Kim M, Na DL, Jeon BS. Magnetic resonance reflects the pathological evolution of Wernicke encephalopathy. J Neuroimaging 2001; 11: 406-11.
11. Weidauer S, Nichtweiss M, Lanfermann H, Zanella FE. Wernicke encephalopathy: MR findings and clinical presentation. Eur Radiol 2003; 13: 1001-9.

ความผิดปกติของสมองชนิด Wernicke: รายงานผู้ป่วย 1 ราย

วีรจิตต์ โชติมงคล, ปณิตา ลิมปะวัฒนะ

รายงานผู้ป่วยชายไทย อายุ 53 ปี มีประวัติดื่มสุราเป็นเวลานาน มาด้วยอาการสับสน ตาซ้ายกลอกขึ้นบนไม่ได้ และเดินเซมา 5 วัน ผลการตรวจ MRI ของสมอง พบลักษณะ non-enhanced low signal intensity on T1WI และ high signal intensity on T2WI กระจายทั่ว ๆ ไปบริเวณ basal ganglia และ thalamus ทั้งสองข้าง, midbrain, pons และรอบ ๆ เวนตริเคิล ร่วมกับสมองและ mamillary bodies ฝ่อ ผู้ป่วยได้รับการรักษาด้วยการฉีดวิตามินบี 1 ทางหลอดเลือดดำ 100 มิลลิกรัมต่อวัน พบว่าความผิดปรกติที่ตาซ้ายดีขึ้นเป็นปกติในวันที่ 8 ของการรักษา ความรู้สึกตัวและอาการเดินเซค่อย ๆ ดีขึ้น