A non-alcoholic case of Wernicke's encephalopathy

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Summary

Wernicke's encephalopathy is an acute neurological disease caused by thiamine deficiency. Alcoholism is the most important risk factor. Recently, non-alcoholic cases are increasingly being recognised. Inadequate dietary intake to meet the metabolic demand or body loss of thiamine is the general explanation. We report a Chinese man who habitually consumed excessive amount of soda water and whose Wernicke's encephalopathy developed after a concurrent pneumonia. Treatment with thiamine achieved dramatic clinical and radiological improvements. Physicians should be aware of this treatable condition among non-alcoholic patients who are eating a traditional Chinese diet.

摘要

魏尼克腦病是維生素B1缺乏所引發的急性腦神經系統疾病。過量飲酒是最重要的危險因素。近年來，我們開始對非酒精性的病例有所認識。其一般成因是由於每日攝入維他命B1不足以滿足身體新陳代謝的需要，或者維他命B1大量流失。有一位華裔男性，有飲用過多汽水的習慣，他在患上肺炎後，出現魏尼克腦病。接受維他命B1治療後，臨床症狀和腦部造影都有明顯的改善。醫生應留意魏尼克腦病不僅限於飲酒過量者，也可以發生在有中國傳統飲食習慣的人士。

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Introduction

Wernicke's encephalopathy should be suspected in chronic alcoholics presenting with relevant neurological symptoms. Early diagnosis and immediate treatment with thiamine are required to minimise the long-term neurological sequelae. A non-alcoholic Chinese patient is reported to illustrate the clinico-radiological features and to alert physicians to a possible association with excessive consumption of soda water and/or traditional Chinese diet.

Case report

A 66-year-old retired painter enjoyed good past health. He is a heavy smoker and a life-long non-drinker. He habitually consumed four to six cans of soda water every day for several years and was eating a traditional Chinese diet. The latter consisted mainly of polished rice, fresh fish, and vegetables.

He presented with drowsiness, listlessness, confused speech, and unsteady gait. These were preceded by fever, cough, dizziness, and anorexia for one week during which he ate less food but drank the same amount of soda water.

Physical examination revealed drowsiness, absence of spontaneous speech, and slow response to verbal communication. He was disoriented in time and place with no insight to his illness. His body weight was 70 kg; blood pressure was 139/81 mmHg, pulse rate was 105 per min, and body temperature was 37.5°C. His neck was supple to passive movements. Bilateral partial ptosis was present, which was worse on the left side. Horizontal eye movements were impossible, and multi-directional nystagmus was elicited upon vertical gaze. Pupils were 2 mm in diameter with sluggish response to light. Fundoscopy was normal. His limb power was full, and sensations were grossly intact. The heel-shin test was clumsy, but other cerebellar signs were not detected. The tendon reflexes were markedly depressed. Plantar responses were down-going bilaterally. Apart from mild consolidation at the left lung base, examination of other systems was unremarkable.

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Wernicke's encephalopathy was suspected, and parenteral thiamine 100 mg daily was given for three days together with antibiotics for his pneumonia. He had dramatic clinical improvement in the next 24 hours with return of alertness and horizontal gaze as well as reduction of nystagmus. Walking became unaided in the next few days; follow-up neurological examination revealed horizontal nystagmus only. He was maintained on oral thiamine 50 mg and a tablet of vitamin B complex daily. Subsequently, memory impairment and confabulation became evident to reveal Korsakoff syndrome.

Regarding investigations, an urgent computed tomography (CT) scan of the brain was normal as well as the complete blood picture and serum electrolytes. Thiamine deficiency was confirmed by the low RBC transketolase activity and an exaggerated ditto-thiamine pyrophosphate effect. These abnormalities were gradually normalised after 4 weeks of thiamine supplementation. Lumbar puncture revealed normal results of cerebrospinal fluid (CSF) analysis. EEG was normal, and nerve conduction studies showed mild peripheral neuropathy.

Magnetic resonance imaging (MRI) of the brain revealed extensive bilateral and symmetrically distributed T2-weighted hyperintensities around the third ventricle, cerebral aqueduct, mammillary bodies, inferior colliculi and posterior medulla without significant contrast enhancement. These are characteristic radiological changes seen in Wernicke's encephalopathy. There was no atrophy of the mamillary bodies, cerebral cortex or cerebellum; ventricles were not dilated. Follow-up MRI of the brain performed one month later showed significant recovery of these radiological abnormalities (**Figures A-F**).

Discussion

Thiamine diphosphate is a coenzyme for several important metabolic pathways. Clinical features of thiamine deficiency reflect impaired enzymatic activities leading to accumulation of the substrates and deficiency of the end-products. Thiamine (vitamin B1) is present in a wide variety of food but largely absent from fat, cassava, refined sugar, and machine-milled rice. In addition, thiamine is rapidly destroyed during cooking at 100°C or above. Furthermore, thiaminases, the inactivating enzymes, are present in fresh fish, clams, shrimps, mussels, and colonic bacteria. Thus, the traditional Chinese diet, which consists of mainly polished rice, steamed fish and over-cooked vegetables, is relatively deficient in thiamine.

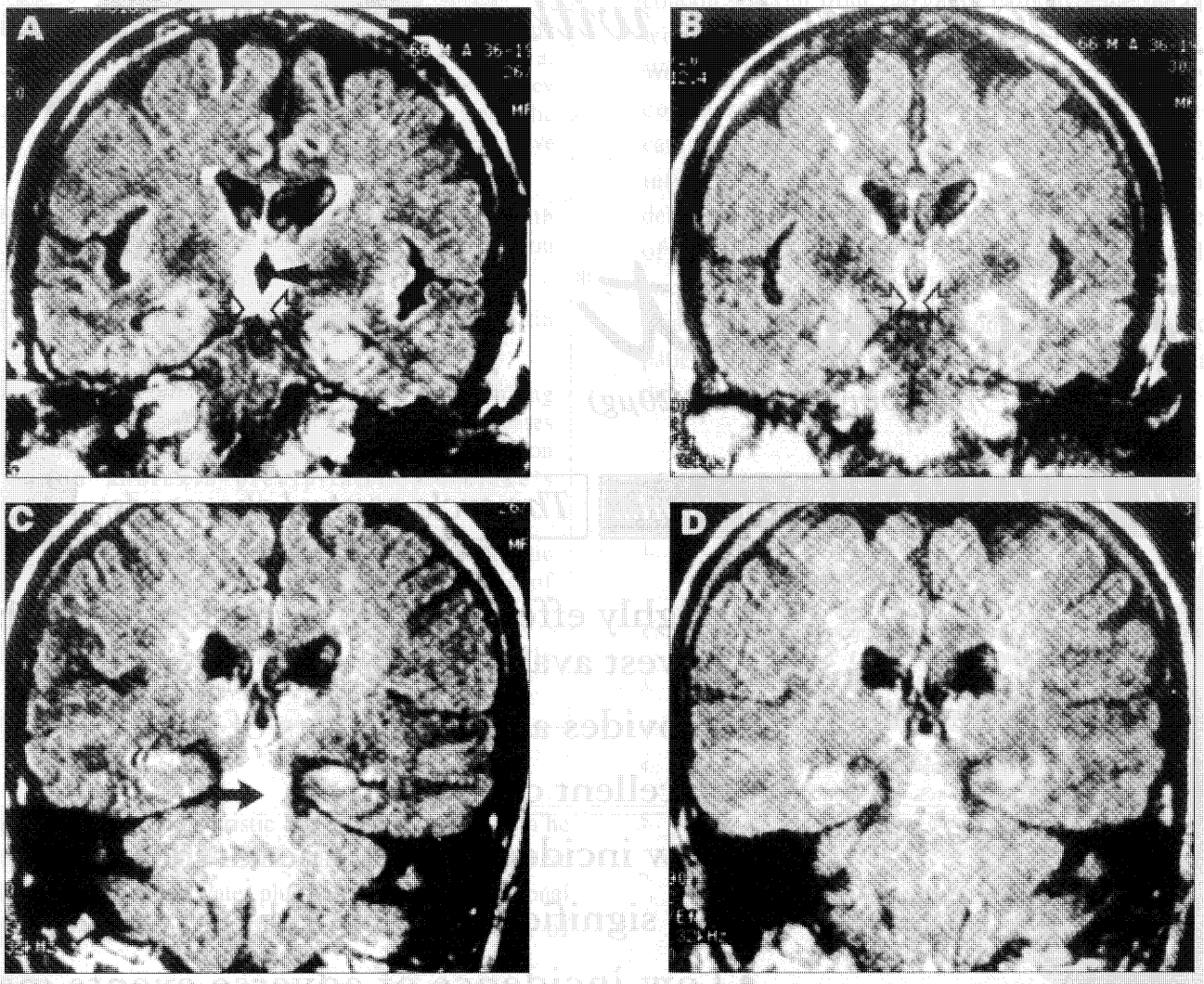
The body demand of thiamine is greatly increased in the following circumstances: carbohydrates being the major energy source, pregnancy, lactation, thyrotoxicosis, and concurrent infections. In addition, body loss of thiamine can be enhanced by the use of diuretics, dialysis, chronic diarrhoea, and malabsorption states. As our body store of the vitamin is small, thiamine depletion and symptoms of deficiency can occur as early as two weeks after experimental institution of a thiamine free diet.

A combination of the above mechanisms accounts for the high risk of developing Wernicke's encephalopathy in chronic alcoholics, although alcohol has neurotoxic effects. Recently, non-alcoholic cases are increasingly reported. Patients with lymphoma or bone tumour on chemotherapy and patients undergoing bone marrow transplantation are liable to develop thiamine deficiency especially if they are receiving total parenteral nutrition.[1] Magnesium depletion (which can be due to diuretics) may cause Wernicke's encephalopathy, because magnesium is an essential cofactor in the production of active thiamine diphosphate and triphosphate esters.[2] Genetic factors have also been postulated to explain an individual's susceptibility to the disease.[3,4]

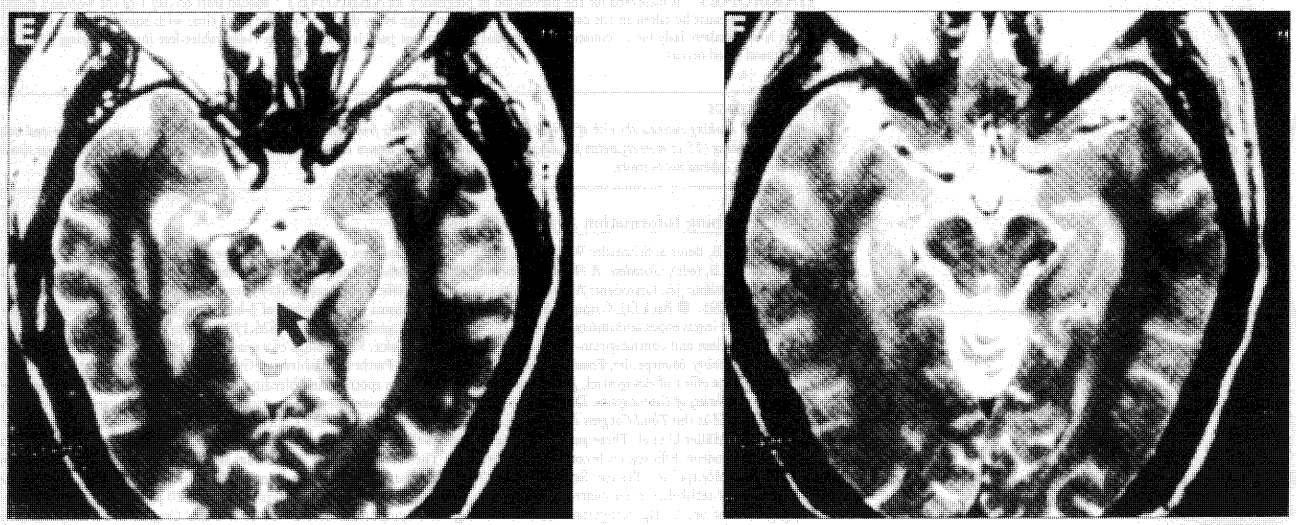
The diagnosis of Wenicke's encephalopathy is essentially clinical, although biochemical assay of RBC transketolase activity and ditto-thiamine pyrophosphate effect can confirm thiamine deficiency. A therapeutic trial of parental thiamine is probably the most useful clinical strategy. Recently, MRI of the brain is increasingly used to reveal the characteristic radiological features of acute Wernicke's encephalopathy.[5] In one MRI study, the sensitivity and specificity are 53% and 93%, respectively.[6] The significance of mamillary body atrophy with or without gadolinium enhancement remains controversial; these radiological features as well as generalised cerebral and cerebellar atrophy are commonly present in chronic alcoholics. CT scan of the brain is generally not helpful.

In our patient, the diagnosis of Wernicke's encephalopathy was based upon a combination of typical clinical features, dramatic response to thiamine replacement, biochemical confirmation of thiamine

Figures A, B, C and D: MRI of the brain showing extensive fluid-attenuation-inversion-recovery



Figures E and F: T2-weighted hyperintensities (E and F) around the third ventricle (long arrow), mamillary bodies (open arrow heads) and periaqueductal regions of the midbrain (short arrows), at baseline (A, C and E) and resolution of these abnormal signals one month after thiamine replacement (B, D and F)



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Key messages

1. Wernicke's encephalopathy is characterised by acute onset of mental disturbance, opthalmoplegia, nystagmus, and ataxic gait. Thiamine deficiency from inadequate dietary intake to meet the metabolic demand and/or body loss of thiamine is the causative mechanism.
2. Early diagnosis and immediate treatment with thiamine are required to minimise the long-term neurological sequelae.
3. Traditional Chinese diet is relatively deficient in thiamine.
4. Excessive consumption of soda water drinking constitutes a high carbohydrate intake and increases the body demand for thiamine. Concurrent infection and anorexia precipitated Wernicke's encephalopathy in this case.
5. Magnetic resonance imaging reveals characteristic changes and can assist in the diagnosis of Wernicke's encephalopathy.
6. Physicians should be aware of this treatable condition among non-alcoholic patients.

deficiency, and characteristic MRI findings. Although he is a life-long non-drinker, the unusual habit of excessive consumption of soda water plus an unbalanced traditional Chinese diet probably provided a background of subclinical thiamine deficiency. Six cans of soda water contain 960 Cal from 240 gm of carbohydrate; the latter would increase the body demand for thiamine. Wernicke's encephalopathy was then precipitated by the concurrent pneumonia, anorexia and continual carbohydrate load; the situation is analogous to glucose infusion in alcoholics. No other cause of thiamine deficiency was identified. This is the first reported case of Wernicke's encephalopathy possibly related to excessive soda water drinking. Physicians should be aware of this treatable condition among non-alcoholic patients who are eating traditional Chinese diet especially during a concurrent illness. ■

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