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LETTER TO THE EDITORS

Non-alcoholic Wernicke encephalopathy presenting as bilateral hearing loss: a case report

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Dear Sirs,

Wernicke encephalopathy (WE) is a syndrome caused by thiamine deficiency whose main clinical features are ophthalmoplegia, ataxia, and altered mental state. This symptom triad can be incomplete, especially in the first few weeks from the onset [1]. WE is typically described as complication of a long-standing history of alcohol abuse, but there is an increase of non-alcoholic cases, mainly due to iatrogenic causes [2].

Here, we report a case of iatrogenic WE in a patient presenting with sudden bilateral deafness 2 months after bariatric surgery.

A 27-year-old-man was admitted to the emergency room (ER) with rapidly progressing bilateral hearing loss followed by balance disturbance and diplopia. Bilateral hypoacusis started few days before and was initially misdiagnosed as otitis media. His relatives reported sleeve gastrectomy approximately 2 months before, followed by recurrent vomiting, and poor compliance with supplementation dietary prescribed after surgery.

General examination was unremarkable, except for Body Mass Index > 40 Kg/mq, indicating severe obesity. On neurological examination, he appeared fully conscious and fairly oriented, but apathy and subtle attention deficit could be noticed. He was unable to understand spoken words, unless with shouting. Cranial nerve examination revealed normal pupillary reaction, but he had impaired bilateral horizontal gaze with end-gaze nystagmus on the left side and bilaterally abnormal horizontal head impulse tests, in absence of upbeat or downbeat nystagmus.

The rest of cranial nerve examination, as well as the motor function, were unremarkable, except for diffuse reduction of deep tendon reflexes with absent Babinski sign. He was not able to stand upright due to severe ataxia, although limb dysmetria was absent. Brain magnetic resonance imaging (MRI) showed symmetric hyperintensities on T2-weighted sequences in bilateral postero-medial thalamus, hypothalamus and mammillary bodies, these latter with contrast-enhancement (Fig. 1a); cochlear and vestibular nuclei and cerebello-pontine angle were spared (Fig. 2). A moderate (30–50 dB at higher frequency, 2000–8000 Hz)-to-severe (80–90 dB at lower frequency, 125–1000 Hz) sensorineural hearing loss was confirmed by reverse-slope audiogram (Fig. 3). Brainstem auditory-evoked responses (BAERs) showed a bilateral prolongation of I-III interval (Fig. 4) and electroneurography supported a peripheral involvement of lower limb nerves.

Vitamin deficit was hypothesized on the basis of clinical history of recent bariatric surgery followed by vomiting, progressive neurological symptoms with acute onset, and MRI picture. Intramuscular thiamine 200 mg thrice daily was then immediately started, even without determination of serum thiamine level (not available in our hospital). Routine laboratory investigations were unremarkable, including red blood cells' indices and Vitamin B12 dosage. His symptoms (hypoacusis, nystagmus and ataxia) began to ameliorate rapidly with thiamine supplementation, and brain abnormalities disappeared almost completely after 2 weeks (Fig. 1b). He was then discharged to neurorehabilitation unit approximately 3 weeks after his admission to the ER.

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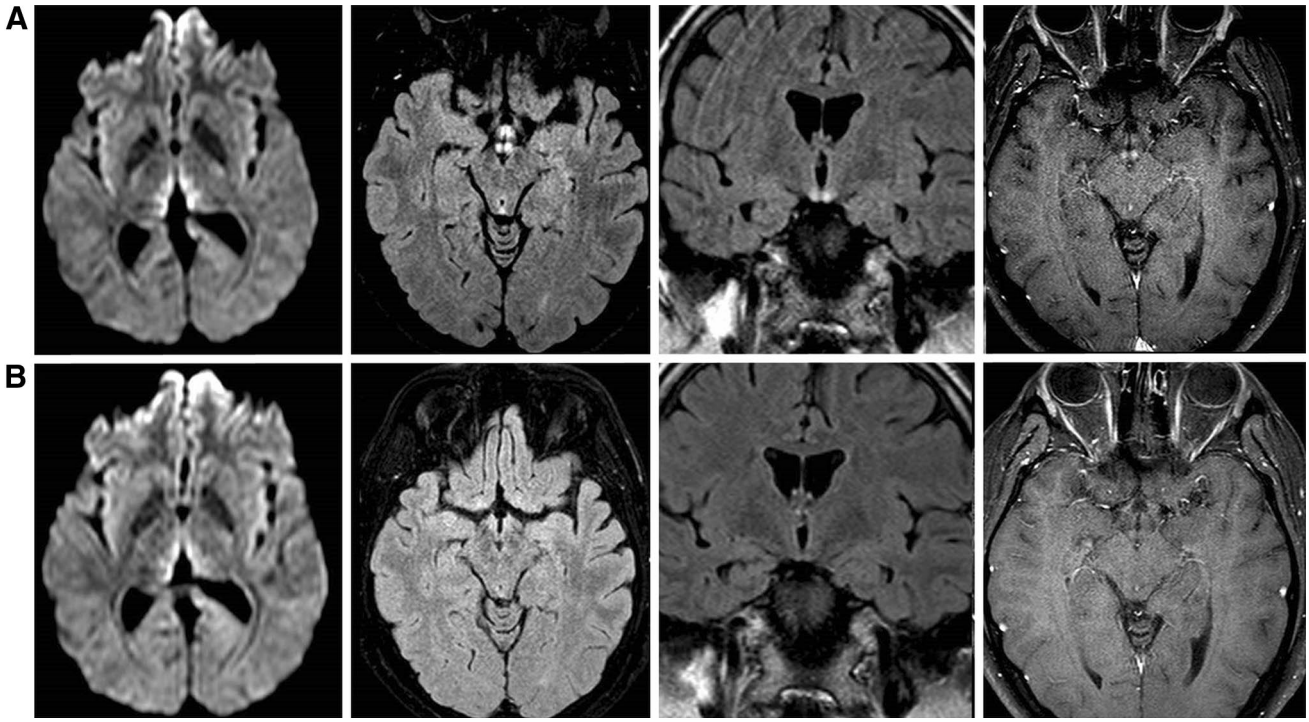

Fig. 1 Magnetic Resonance Imaging (MRI) of the brain before (**a**) and after (**b**) thiamine supplementation (sequences from left to right: axial diffusion-weighted; axial fluid-attenuated inversion recovery; coronal fluid-attenuated inversion recovery; axial post-contrast T1-weighted)

Fig. 2 Axial T2-weighted slightly off plane images showing normal-appearing restiform bodies, containing the cochlear and vestibular nuclei (**a**) and right and left normal cerebello-pontine angles and internal auditory canals (**b**)

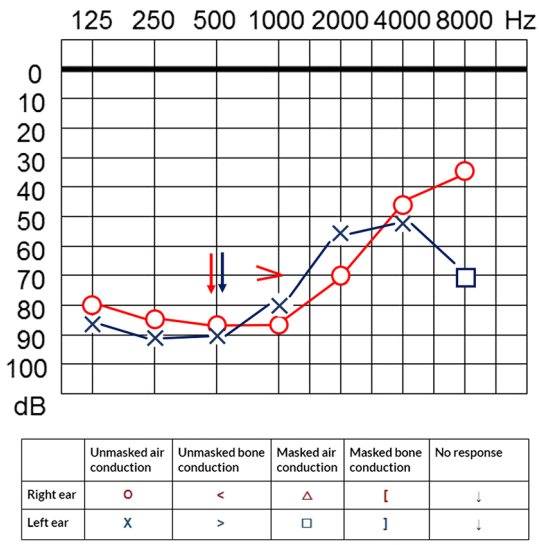

Fig. 3 Reverse-slope audiogram obtained prior to initiation of thiamine supplementation showing sensorineural hearing loss bilaterally

Hearing loss is an uncommon presentation of WE, mainly reported in non-alcoholic cases [3]. Our case points out that initial symptoms of WE are often not recognized as such, especially in the presence of atypical features that are more common in non-alcoholic WE [4]. Although the classic triad was incomplete in our patient, one can argue

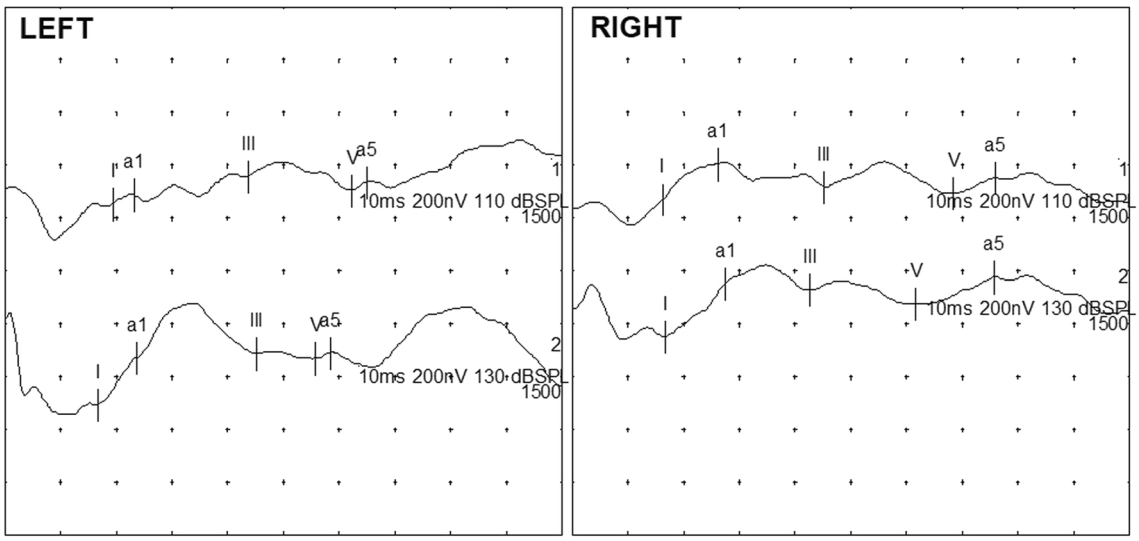

Fig. 4 Brainstem auditory-evoked responses: prolonged I-III interpeak latencies bilaterally elicited by auditory stimulus

that the concomitant deafness prevented the detection of a subtle memory impairment. On the other hand, mental status change has been reported to be unlikely in younger bariatric patients developing WE [5]. Anyway, the operational diagnostic criteria for WE were fulfilled in our case [6].

WE-related deafness has been attributed to involvement of geniculate nuclei of thalamus [7], but our case suggests instead a peripheral damage to acoustic nerves as revealed by BAERs and the sparing of cochlear nuclei and inferior colliculi at brain MRI. Peripheral neuropathy, detected also in our case, is sometimes accompanied by hidden hearing loss [8] and is often reported in patients with non-alcoholic WE [9]. Moreover, basic research studies showed an auditory neuropathy phenotype in mice lacking high-affinity thiamine transporter, thus suggesting damage to the sensory inner hair cells, their ribbon synapses or spiral ganglion neurons caused by thiamine deficiency [10].

In conclusion, WE should also be taken into account in case sudden hypoacusis occurs 4–12 weeks after bariatric surgery, especially if followed by recurrent vomiting that represents the most relevant precursor of WE due to depletion of thiamine stores in these patients [11]. Accurate vestibular bedside testing should also be performed in any case of unexplained acute or subacute hearing loss, even in absence of encephalopathy [12, 13], to confirm suspected WE and to start thiamine supplementation promptly.

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Compliance with ethical standards

Ethical approval The patient provided informed oral and written consent in accordance with specific national laws and the ethics standards laid down in the 1964 Declaration of Helsinki and its later amendments.

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