Atypical imaging manifestations in non-alcoholic Wernicke's encephalopathy: a potentially reversible neurological condition not to be missed

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An 18-year-old female with good past health was diagnosed with right tibial osteosarcoma in February 2019. She underwent wide excision of the right proximal tibia and distal femur with total knee replacement. Postoperatively, her adjuvant chemotherapy was complicated by multiple episodes of opportunistic infection, acute renal impairment due to drug toxicity and electrolyte disturbance. She was hospitalised for >6 months with suboptimal oral intake.

Over the course of a week, the patient had two episodes of seizure. Her general consciousness deteriorated acutely to a Glasgow Coma Scale score of 8/15 (E4V1M3). Physical examination revealed decorticate posture, generalised flaccidity and areflexia. Serum sodium level and urea were markedly elevated (154 mmol/L and 17.0 mmol/L, respectively), in keeping with hypernatraemic dehydration. Electroencephalogram showed diffuse slow-wave encephalopathy. Her Glasgow Coma Scale score did not improve following correction of hypernatraemia.

Magnetic resonance imaging of the brain revealed an abnormal high T2-weighted signal and restricted diffusion in bilateral frontal lobe cortices, dorsomedial thalami, periaqueductal grey, tectal plate of the midbrain and mammillary bodies (Fig 1a-d). Based on these findings, the patient was diagnosed with Wernicke's encephalopathy and high-dose intravenous thiamine (vitamin B1) was initiated. Although her level of consciousness improved rapidly, there was poor recovery of limb power. Follow-up magnetic resonance imaging of the brain demonstrated cortical laminar necrosis and haemorrhage at bilateral frontal cortices (Figs 1e and 2). After 2 years of intensive rehabilitation, she regained most of her upper limb power, but lower limb power remained impaired.

Wernicke's encephalopathy is an acute neurological syndrome caused by depletion of intracellular thiamine in neurons that is essential for production of neurotransmitters. The bodily reserve of thiamine in a healthy individual is exhausted within 4 to 6 weeks in the absence of dietary thiamine.[1] Wernicke's encephalopathy is most commonly associated with chronic alcoholism but can result from any condition that causes malnutrition or malabsorption.[1] The classic clinical triad consists of confusion, ataxia and ophthalmoplegia, although only a small proportion of patients exhibits all three.[2] Left untreated, Wernicke's encephalopathy carries significant neurological morbidity and death. The condition is potentially reversible if recognised and treated early with intravenous thiamine replacement.

Classic imaging features of alcohol-associated Wernicke's encephalopathy include abnormal signal involving deep periventricular and periaqueductal grey matter in basal ganglia and brainstem, most notably in the mamillary bodies.[3] Atypical findings are more frequently seen in non-alcoholic Wernicke's encephalopathy. These include abnormal signal in other locations such as the cerebral cortex, splenium, caudate nuclei, red nuclei, cranial nerve nuclei, cerebellum and vermis.[4] Further progression to cortical laminar necrosis and haemorrhage, as seen in our case, is rare and associated with a poor prognosis due to irreversible neurological damage.[5]

In patients with a poor nutritional state who present with reduced consciousness, a high index of clinical suspicion and prompt imaging are important to establish the diagnosis of Wernicke's encephalopathy. Atypical imaging manifestations are more commonly seen in non-alcoholic Wernicke's encephalopathy. Timely diagnosis is crucial since the neurological impairment is potentially reversible with intravenous thiamine replacement therapy.

Author contributions

Concept or design: CCY Chan, KKF Fung.
Acquisition of data: CCY Chan, KKF Fung.
Analysis or interpretation of data: All authors.
Drafting of the manuscript: CCY Chan, KKF Fung.
Critical revision of the manuscript for important intellectual content: All authors.

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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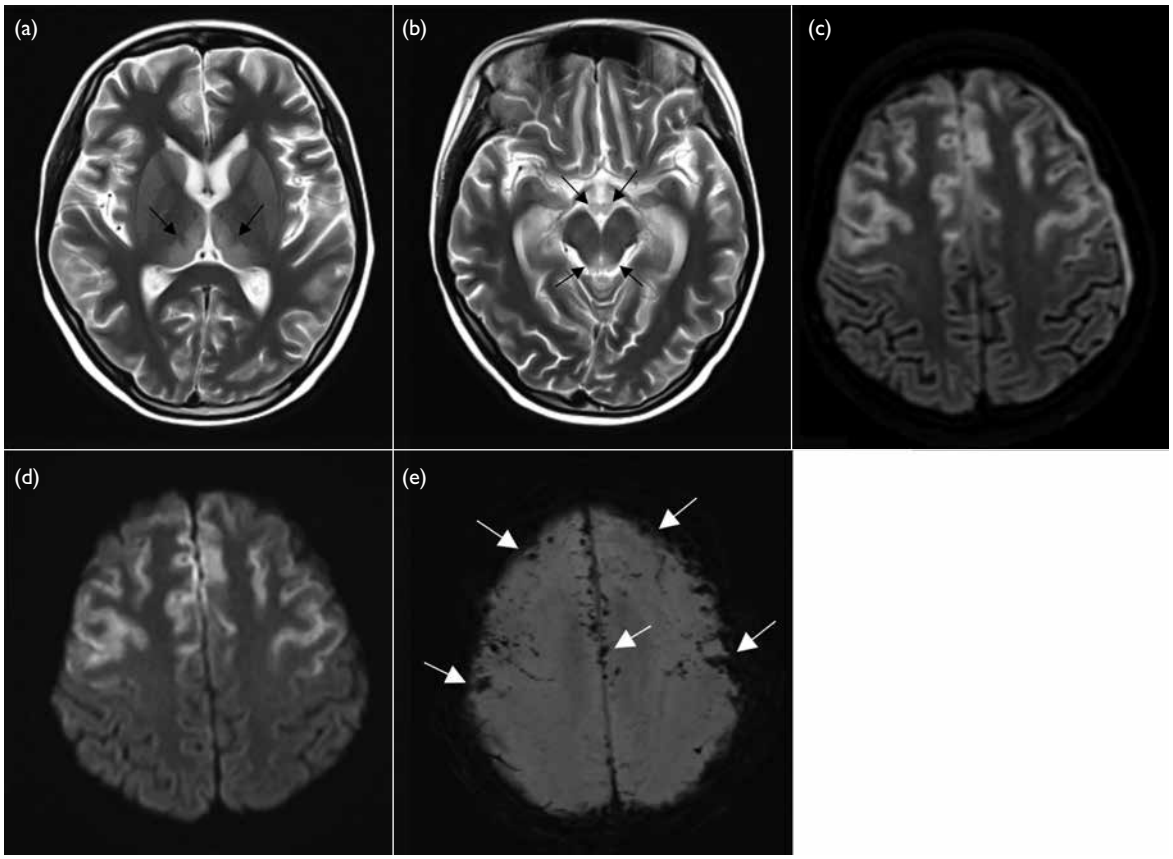

FIG 1. Magnetic resonance imaging (MRI) of the brain demonstrated atypical imaging features in addition to the classic findings in Wernicke's encephalopathy. Axial T2-weighted MRI showing abnormal symmetrical T2-weighted signals in bilateral dorsomedial thalami (a) and periaqueductal grey, tectal plate of the midbrain and mammillary bodies (b), which are typically seen in Wernicke's encephalopathy (black arrows). (c) Axial fluid-attenuated inversion recovery MRI showing abnormal high signal involving cortices of bilateral frontal lobes. (d) Diffusion-weighted imaging (*b* value=1000 s/mm^2) showing restricted diffusion in bilateral frontal lobe cortices. (e) Susceptibility-weighted imaging showed blooming artifacts in bilateral frontal lobe cortices, indicating microhaemorrhage (white arrows). The involvement of frontal lobe cortices with haemorrhage is an atypical imaging feature and more commonly seen in non-alcoholic Wernicke's encephalopathy

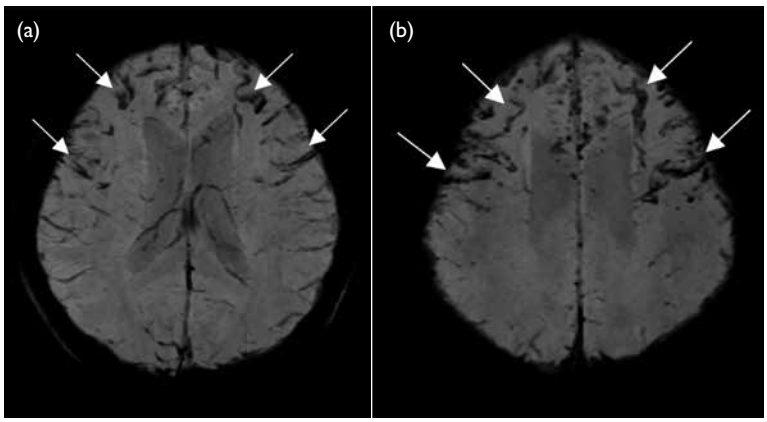

FIG 2. (a, b) Susceptibility-weighted sequences in follow-up magnetic resonance imaging of the brain revealed curvilinear blooming artifacts at cortices of bilateral frontal lobes, indicating development of cortical laminar necrosis (white arrows). This is associated with poor neurological prognosis due to irreversible damage

Ethics approval

The patient was treated in accordance with the Declaration of Helsinki. Informed consent was obtained from the patient for all treatments and procedures, and consent for publication.

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