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Wernicke's encephalopathy with pinpoint pupils and diplopia

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Abstract

This case report presents the clinical findings of a female patient diagnosed with Wernicke's encephalopathy, characterized by pinpoint pupils. While pupillary changes can occur in Wernicke's encephalopathy, the presence of pinpoint pupils is exceedingly rare. In this report, we aim to document and discuss this unusual presentation, as well as speculate on the potential mechanisms underlying this atypical manifestation of the disease.

Keywords Wernicke's encephalopathy, Pinpoint pupils, Diplopia

Introduction

Wernicke's encephalopathy is a metabolic disorder of the central nervous system caused by vitamin B1 deficiency. Its etiology can be divided into: alcohol-related (chronic alcoholism) and non-alcohol-related (such as vomiting in pregnancy, gastrointestinal surgery, frequent vomiting, chronic diarrhoea, cancer chemotherapy, dialysis, etc.).This disease often symmetrically involves the mammillary bodies, thalamus, tissue around the third ventricle, periaqueductal gray matter, and other areas of rapid glucose metabolism and sensitivity to thiamine deficiency, with the mammillary bodies being the most vulnerable site [1, 2]. Typical clinical manifestations are the 'triad signs' - oculomotor disorders, ataxia, and mental consciousness disorders [3]. However, in our case report, the symptoms and the MRI of the patient were not obvious upon initial admission to the hospital.She was even misdiagnosed with Miller-Fisher syndrome and exhibited rare pinpoint pupils and diplopia.

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Case presentation

A female patient in her 50s received parenteral nutrition in the gastrointestinal surgery department for one month due to intestinal obstruction, and then developed dizziness, ocular movement disorders, bilateral pinpoint pupils, diplopia, photophobia, repeated refractory hiccups, and unsteady gait. Upon physical examination, it was revealed that the patient exhibited limited convergence reflex, adduction, abduction, upward and downward gaze of both eyes. However, the light reflex was found to be normal (Video 1). And the patient did not have ptosis or significant nystagmus.The Romberg sign was positive and tendon reflexes were decreased. Additionally, the patient's speech comprehension ability and reaction ability were dull. The rest of the physical examination was normal.In addition, the patient denied organophosphorus or choline poisoning. The patient also denied congenital miosis and other ophthalmological disorders. Moreover, the patient's pupils size and light reflex were normal when she was first admitted to the gastrointestinal surgery department, and her family did not notice any similar pupil abnormalities.

Initially, the patient was diagnosed with Miller-Fisher syndrome in the gastrointestinal surgery department. On March 8th, brain Magnetic resonance imaging (MRI) and electromyogram (EMG) were normal (Fig. 1 March 8th).

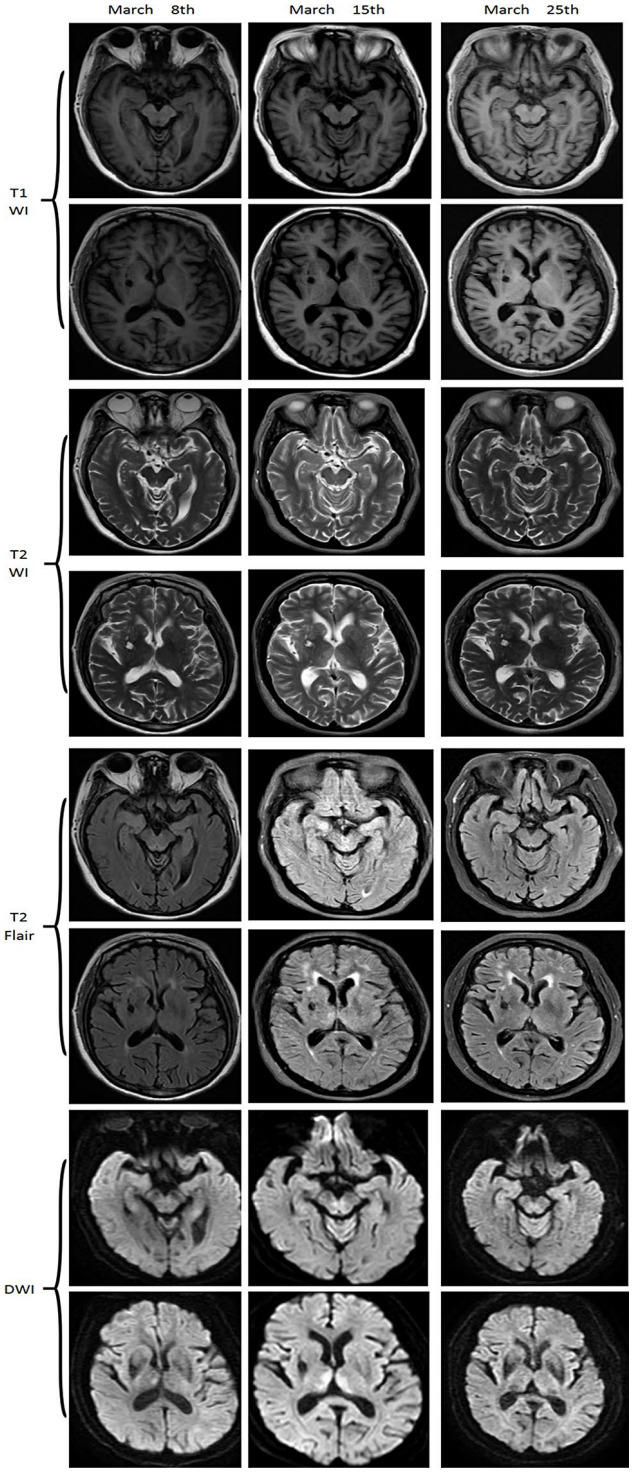

Fig. 1 Brain MRI changes of the patient Brain MRI on March 8th did not show significant lesions on T1WI, T2WI, T2Flair and DWI. Brain MRI on March 15th showed hyper signal lesions in the midbrain aqueduct and bilateral thalamus on T2WI, DWI and T2flair. After treatment, brain MRI on March 25th showed smaller hyper signal lesions in the midbrain aqueduct and bilateral thalamus on T2WI, DWI and T2flair

Lumbar puncture revealed no obvious protein-cell separation phenomenon, and the examination of rheumatism series showed no obvious abnormality.The patient did not receive vitamin B1 treatment during her hospitalisation in gastrointestinal surgery department. The patient was referred to the Department of Neurology on 11 March, where she was initially considered to have Wernicke's encephalopathy and was treated with vitamin B1 immediately after blood tests for vitamin B1 levels were drawn. And on March 15th, the patient's brain MRI showed hyper signal lesions in bilateral thalamus and aqueduct of midbrain (Fig. 1 March 15th). The blood vitamin B1 levels were low at 41.5 mmol/L (normal range: 70–180 mmol/L), and Aquaporin-4 (AQP-4) antibody presented no discernible abnormalities. As a result, Thus, the final diagnosis was Wernicke's encephalopathy rather than Miller-Fisher syndrome and treatment with vitamin B1 was continued.

After receiving vitamin B1 treatment, the patient showed significant improvement in repeated hiccups, pupil size, diplopia, ophthalmoplegia and ocular movement disorders compared to admission. On March 25th, the completed MRI of the brain revealed improvements in the high-signal lesions in the bilateral thalamus and the cerebral aqueduct of the midbrain on T2-weighted image(T2WI), diffusion weighted imaging (DWI), and T2 fluid attenuated inversion recovery (Flair) compared to before. This indicates a positive development in the patient's condition. Subsequent outpatient follow-up indicated that the patient's vitamin B1 levels had returned to normal. Although the patient's pinpoint pupils had improved, they had not returned to their original size. Additionally, eye movements and diplopia were better than before.

Discussion and conclusion

We report a case of Wernicke's encephalopathy with marked pinpoint pupils and diplopia in addition to the typical features of Wernicke's encephalopathy. The initial MRI of the brain did not reveal obvious lesions [4]. And the patient was initially diagnosed with Miller-fisher syndrome.The lumbar puncture was performed without obvious abnormality. However, as the patient's condition progressed, the lesions were observed on in the MRI. Following treatment with vitamin B1, the lesions gradually improved, but the pinpoint pupils did not return to its original size. After discharge, the patient was followed up in the outpatient clinic and was significantly better than before.While pupillary changes can occur in Wernicke's encephalopathy, the occurrence of Wernicke's encephalopathy with pinpoint pupils is extremely rare [5]. Numerous previous literature has shown that changes in pupil size are closely related to damage to sympathetic pathways [6–8]. Damage to different sites

of the sympathetic pathway will result in different clinical symptoms.The sympathetic pathway originates in the hypothalamus, descends via the brainstem, and leaving the spinal cord and ascending within the sympathetic chain. The neurons travel first in the carotid artery wall, join the abducens nerve in the cavernous sinus, and thereafter join the ophthalmic nerve, pass the ciliary ganglion, and reach the pupillary dilator muscle [9–11]. In addition, Wernicke's encephalopathy typically involves the hypothalamus, periventricular tissues of the third ventricle, and tissues surrounding the midbrain aqueduct. Moreover, sympathetic fibres from the hypothalamus and insula pass down through the periventricular tissues of the third ventricle, where the pupil-dilating centre of the sympathetic fibres and the patient's lesion are located.The patient's sympathetic fibres were selectively affected, resulting in pinpoint pupils. We consider that this is the possible mechanism by which the patient developed this symptom.We also believe that patients with this disease may have normal MRI in the early stages of the disease and that the typical symptoms are a late finding, so clinicians should recognise thiamine depletion in time to enable appropriate treatment and prevention of Wernicke's encephalopathy and full recovery [3, 4].

Abbreviations

MRI	Magnetic resonance imaging
T2WI	T2-weighted image
EMG	Electromyogram
AQP-4	Aquaporin-4
DWI	Diffusion weighted imaging
Flair	Fluid attenuated inversion recovery

Supplementary Information

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Supplementary Material 1: Video 1. The patient had pinpoint pupils in both eyes, normal light reflexes in both eyes, limited inward and outward eye movements in both eyes, and unrestricted upward and downward movements.

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Author contributions
Hongjia Xu: Drafting/revision of the manuscript for content, including medical writing for content. Na Shao: Major role incollecting and analyzing literature. Zhengyu Zhu: Analysis or interpretation of pathogenesis. Pin Wang: Drafting/revision of the manuscript for content, including medical writing for content. Lin Sun: Drafting/revision of the manuscript for content, including medical writing for content. Yingying Xu: Drafting/revision of the manuscript for content, including medical writing for content; Analysis or interpretation of Pathogenesis.

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Data availability
All data and materials used in this study are available from the corresponding author on request.

Declarations

Ethics approval and consent to participate
Written informed consent was obtained from the patient for publication of this case and accompanying images and video. The study is approved by ethics committee of the Second Hospital of Shandong University.

Consent for publication
Written informed consent was obtained from the patient for publication of this case and accompanying images and video.

Disclosure
The authors report no disclosures relevant to the manuscript.

Competing interests
The authors declare no competing interests.

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