Message in the bottle: Wernicke-Korsakoff Syndrome

E O'Driscoll, S Casey, E Sweeney, A Corvin
Department of Psychiatry, Jonathan Swift Clinic, St James's Hospital Dublin 8

Introduction

Wernicke's encephalopathy (WE) is a neurological condition resulting from thiamine deficiency with varied neurocognitive manifestations, typically involving mental state changes, gait and oculomotor dysfunction. (1)

Thiamine deficiency is characteristically associated with severe alcohol use disorder. Although WE primarily affects people who have a thiamine deficiency due to chronic alcoholism, various other causes include severe malnutrition, hyperemesis gravidarum, prolonged parenteral nutrition, malignancies, immunodeficiency syndromes, liver disease, hyperthyroidism, and severe anorexia nervosa.

Chronic alcohol consumption may cause thiamine deficiency due to impaired absorption of thiamine from the intestine, a possible genetic predisposition, inadequate diet, reduced storage of thiamine in the liver, and other nutritional deficiencies. (1)

Due to unclear diagnostic criteria for Wernicke's encephalopathy, it is difficult to diagnose and often goes untreated or undertreated in clinical practice. This lack of detection and treatment is evidenced by the fact that Wernicke's encephalopathy is first diagnosed post-mortem in over 80% of cases. (2)

Diagnostic History of WE

Carl Wernicke 1881

All of the following:

- Confusion
- Ophthalmoplegia
- Ataxia

Caine et al operational Criteria adopted by European Federation of Neurological Services (3)

Patients demonstrating 2 of the 4 following criteria:

- Dietary deficiencies
- Eye signs
- Cerebellar signs
- Altered mental state
- Mild cognitive impairment

Royal College of Physicians (United Kingdom) (4)

Patient with evidence of chronic alcohol misuse plus 1 of the following:

- Acute confusion
- Decreased level of consciousness
- Memory problems
- Ataxia
- Ophthalmoplegia
- Hypothermia
- Hypotension

Case History and Clinical Course

We present the case of a 47-year-old female who was admitted to our inpatient unit via form 10 of the Mental Health Act 2001 with acute behavioural disturbance of unclear aetiology.

This individual had historically attended this service with depressive episodes and a history of alcohol dependence syndrome (ADS) leading to a differential of mania versus behavioural effects of alcohol.

She presented as a bizarre historian; chaotic, disorganized and disorientated. She failed to respond to pharmacological intervention with antipsychotic medications at optimal dosages. Thiamine was initiated given her history of ADS. The patient's mental state failed to stabilise initially and was suggestive of hyperactive delirium in light of ongoing confusion and agitation. Blood tests demonstrated deranged liver function but were otherwise unremarkable.

Collateral from other psychiatric services was obtained indicating multiple previous admissions under psychiatry with a similar presentation leading to a variety of diagnoses including; recurrent depressive disorder, hypomania and most recently factitious disorder.

Results

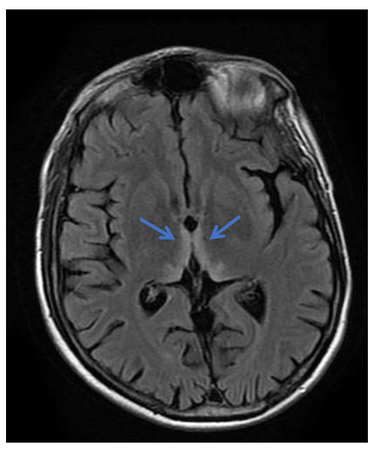
During the most recent admission to St James's Hospital, patient presented with impairment of both anterograde and retrograde memory, however immediate recall was intact.

She demonstrated confabulation. Longitudinal assessment of mental state suggested a diagnosis of Wernicke - Korsakoff syndrome (WKS) which was supported by response to thiamine supplementation.

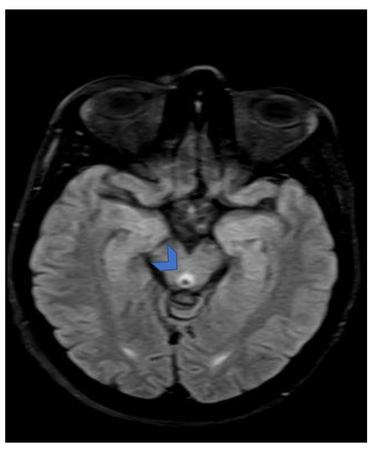
When WKS was suspected, oral thiamine was substituted with parenteral treatment.

MRI revealed white matter changes consistent with chronic small vessel ischaemia, without frontal lobe abnormalities and/or manifestations of WKS.

Typical MRI Findings in WE



Axial FLAIR MRI images demonstrate signal hyperintensity in the dorsomedial thalami and around the third ventricle (arrows), and in the periaqueductal grey matter (arrow head).



High signal is also seen in the mammillary bodies and tectal plate. T1 post contrast and Diffusion-weighted sequences show similar changes. (5)

Conclusion

WE is a medical emergency that is poorly understood and underrecognized. (6)

We present this case to highlight the diverse way in which WE may present which can delay initiation of treatment.

Given the variability in presentations and serious consequences of untreated WE, we recommend a high index of suspicion and a low threshold for early treatment with thiamine.

References

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Declarations
Written consent was obtained from patient concerned in this report. The author thanks the patient's willingness and compliance with participation in this case report.