Atypical Wernicke's syndrome sans encephalopathy with acute bilateral vision loss due to post-chiasmatic optic tract edema

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Abstract

A middle aged male presented with acute bilateral vision loss, 4 weeks after undergoing gastric bypass surgery for gastric carcinoma. He had normal sensorium, fundoscopy, normal pupillary reaction to light, but had mild opthalmoparesis and nystagmus with ataxia. Magnetic resonance imaging of the brain revealed post-chiasmatic optic tract edema along with other classical features of Wernicke's syndrome. Thiamine supplementation leads to complete resolution of clinical as well as imaging findings. In appropriate clinical settings, a high index of suspicion and early treatment are essential for managing Wernicke's syndrome even in patients with atypical clinical and imaging presentation.

Key Words

Gastric surgery, optic tract, thiamine, vision loss, Wernicke's encephalopathy, Wernicke's syndrome

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Introduction

Wernicke's encephalopathy (WE) was first described in 1881 by Carl Wernicke.[1] It presents with the classical triad consisting of mental confusion, ophthalmoplegia and gait ataxia. WE is a serious, but curable neurological disease caused by thiamine deficiency that occurs most commonly in people with alcoholism and its prognosis depends on the commencing time of thiamine supplementation, the most important factor relative to a curative effect.[1-3] Early accurate diagnosis of WE is critical in clinical practice, but patients often do not present with the classic clinical triad. The term Wernicke's syndrome or atypical WE is often used to describe patients who present with atypical symptoms without the classical triad. There are only few reports in the literature describing acute vision loss as a presenting feature of Wernicke's syndrome, with variable neuro-anatomic abnormality as a cause of blindness.[4,5] We describe a patient presenting with acute vision loss in the clinical setting of WE without the classical triad and with rare imaging findings and improvement with thiamine supplementation.

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Case Report

A 69-year-old male presented with intermittent feeling of blurred vision with mild imbalance while walking for 2 days followed by acute loss of vision in both his eyes. He had undergone partial gastrectomy for poorly differentiated adenocarcinoma of the pylorus before 1 month. He had no other health related problems previously. His family history was unremarkable and he had no history of alcohol or tobacco consumption. The immediate post-operative period was unremarkable. Ophthalmologic evaluation revealed total loss of visual acuity with no light appreciation or finger counting with preserved bilateral pupillary reaction to light. There was mild opthalmoparesis with reduced saccadic and pursuit movements toward the sound and patient's own finger movements. There was bilateral horizontal gaze evoked nystagmus with fast component in the direction of gaze. Bilateral optic fundi were normal. He had symmetric trunk and gait ataxia, but no limb ataxia or dysarthria. His alertness, mood, memory, behavior and other cognitive functions were well-preserved. His other systemic and neurologic examination was unremarkable. An urgent plain and contrast magnetic resonance imaging (MRI)

of the brain was done to rule out either of posterior circulation stroke (Bilateral Posterior Cerebral Artery infarct), intracranial metastasis and paraneoplastic neurological dysfunction in view of recently operated carcinoma of stomach.

MRI brain revealed bilateral symmetrical hyper-intensities over the mammillary bodies and periventricular regions of the third ventricle, bilateral inferior colliculi on T2-weighted and fluid attenuated inversion recovery (FLAIR) images [Figure 1a, b, c and d] with homogenous enhancement postcontrast [Figure 1e and f] with no restriction on diffusion-weighted and apparent diffusion coefficient (ADC) images (ruling out ischemia/cytotoxic edema). In addition to these classic findings of WE, focal hyper-intensities were also observed in bilateral post-chiasmatic optic tract on FLAIR images [Figure 1a]. Based on the imaging findings and clinical background of patient, diagnosis of non-alcoholic acute Wernicke's syndrome was considered and treatment with intravenous thiamine hydrochloride (500 mg/d) was promptly started. His visual symptoms significantly improved in 24 h and his vision became completely normal in 72 h. His ataxia improved gradually over the next 3 weeks. He was discharged with the advice to take intravenous thiamine injections once every month to prevent the thiamine deficiency. A follow-up MRI brain carried out after 1 month revealed complete resolution of MR signal intensities [Figure 2a and b].

Discussion

WE usually results from chronic alcohol consumption related malnutrition.[1,3,6] A high index of suspicion for non-alcohol related WE is required in patients with prolonged fasting, parenteral nutrition, long-term intravenous glucose infusion like patients with corrosive ingestion or hyperemesis gravidarum.[2-4] Apart from these, WE can also occur after gastrointestinal surgical procedures carried out for cancer or obesity surgery, including gastro-jejunostomy, total or partial gastrectomy, gastric bypass surgery.[2,3,7,8] In patients with gastric surgery, WE is reported to develop usually weeks or months after the operation. Our patient developed Wernicke's syndrome 1 month after partial gastrectomy with episodes of vomiting, few patients develop the disorder as early as 2 weeks post-operatively and few as late as 20 years after gastrectomy.[1,7,8]

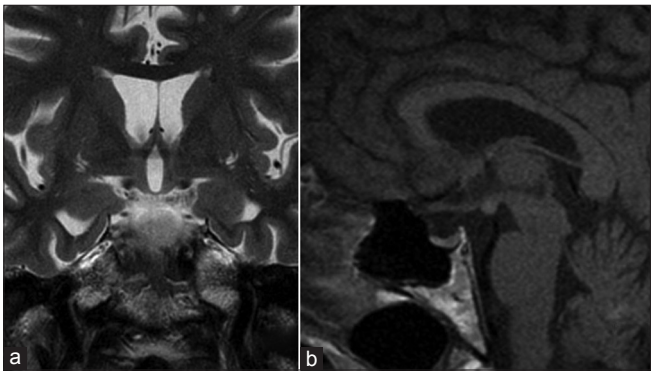

Figure 2: Follow-up T2W coronal image (a) and postcontrast T1W sagittal image (b), shows complete resolution of abnormal signals in mamillary bodies, inferior colliculi and in post-chiasmatic optic tracts

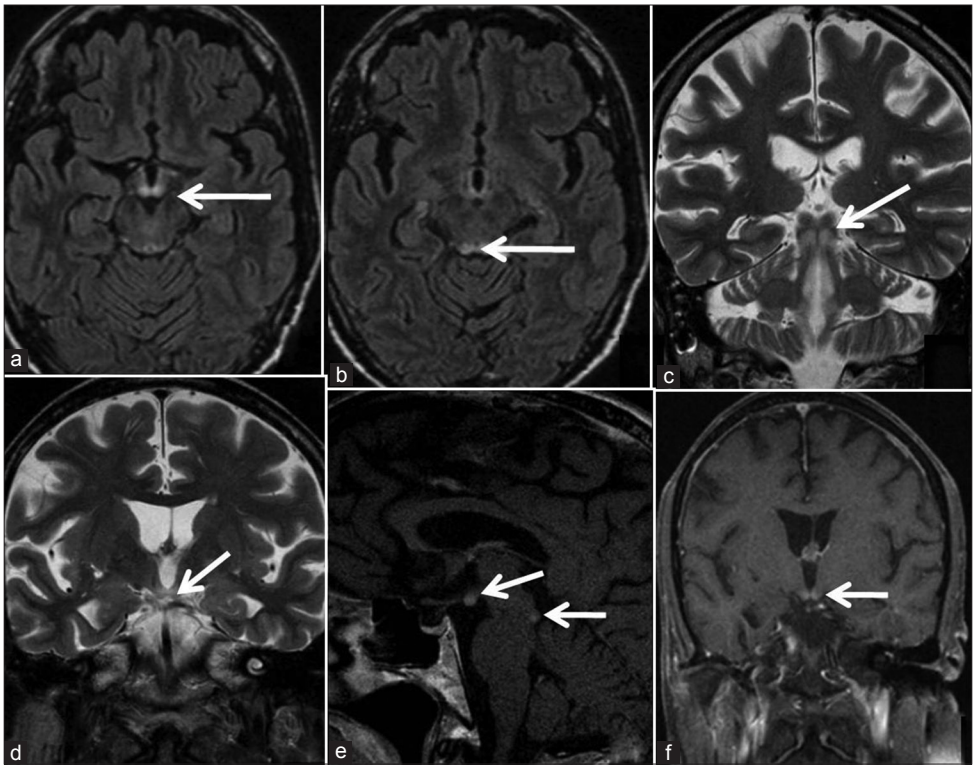

Figure 1: Axial fluid attenuated inversion recovery images (a, b) and coronal T2W images (c, d) show abnormal symmetrical hyper intensities in bilateral post-chiasmatic optic tracts (a), bilateral mamillary bodies (b, d), periaqueductal area (b, d) and in bilateral inferior colliculi (c), Postcontrast T1 sagittal image (e) and coronal T1W image (f), shows homogenous bilateral symmetric enhancement in mamillary bodies and in inferior colliculi

Non-alcoholic WE exhibiting atypical presentation has a high likelihood of being missed initially due to non-specific symptoms, poorly recognized clinical signs as well as unavailability of serum thiamine level testing in most hospitals. In this regard, MRI is currently considered the most valuable method to confirm a diagnosis of WE. The typical findings of MRI are observed in only 58% of WE patients. MRI exhibiting increased T2 signal intensity in the periaqueductal regions of the midbrain and periventricular regions of the thalamus had a sensitivity of 53% and specificity of 93% for the diagnosis of acute WE.[1,9,10] Classical MRI features of WE include areas of increased T2-weighted and FLAIR signals and symmetric areas of contrast enhancement after gadolinium injection in the thalamus, periventricular region of the third ventricle, mamillary bodies, periaqueductal area, tectal region, periventricular gray matter of the fourth ventricle.[3,9,10,11] Restricted diffusion characterized by high signal on diffusion-weighted imaging (DWI) and low signal on ADC maps has occasionally been described in the thalamic region.[12,13] In our patient, MRI did not show any restriction on DWI in the involved structures, but there were hyperintensities on T2, FLAIR images and postcontrast enhancement on T1 images. These findings may be the result of vasogenic edema secondary to metabolic derangement caused by thiamine depletion, which showed complete reversal on repeat imaging after thiamine supplementation. The varied MRI findings in a different case reports in the literature are probably the result of the imaging findings being dependent on the timing of MRI after acute thiamine deficiency as well as the severity of the deficiency, determining the degree of edema in different brain structures.

Our patient developed acute bilateral vision loss of post-chiasmatic localization in the setting of thiamine deficiency and MRI demonstrated bilateral optic tract enhancement, which well-explained the patient's symptoms neuro-anatomically. The presentation of acute vision loss in association with other features of WE has been published in literature before, but in those cases vision loss has been related to the optic disc edema or retinal dysfunction due to intra retinal hemorrhage as a cause of vision loss, which could be related to alcohol toxicity or as a part of WE manifestations.[4,5,13] Our patient had normal fundoscopy, normal pupillary light reflex; thus ruling out optic disc, optic nerve or retinal abnormality as a cause of vision loss and so our patient's presentation with visual loss due to post-chiasmatic bilateral optic tract involvement is quite unique in comparison to other described cases.

Conclusion

We conclude that the complex clinical picture with initial acute vision loss blindness without any confusion or encephalopathy can be an unusual presentation of Wernicke's syndrome. The finding of post-chiasmatic optic tract edema in association with vision loss as the presenting feature of WE is rare. These atypical clinical and neuroimaging findings should not deter the clinician from making the diagnosis of acute Wernicke's syndrome even in non-alcoholics. There should be a high index of suspicion of Wernicke's syndrome in specific patient populations such as alcoholism, post-gastric surgery. Early recognition of atypical clinical presentations including visual loss, adequate utilization of advances in MRI and prompt administration of therapy can improve the prognosis in this potentially reversible neurological emergency.

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