Ocular manifestations in a case of Wernicke’s encephalopathy due to hyperemesis gravidarum

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Abstract:
Wernicke’s Encephalopathy (WE) is a potentially devastating disorder, which if not diagnosed and treated at the earliest, could lead to dangerous neurological and ophthalmological complications. We report the ocular manifestations of WE in a pregnant woman with hyperemesis gravidarum.

Keywords:
Hyperemesis gravidarum, optic disc edema, retinal hemorrhages, thiamine, Wernicke’s encephalopathy

Introduction
Wernicke’s encephalopathy (WE) is a neurological disorder characterized by the classic triad of encephalopathy, ophthalmoplegia, and/or nystagmus and ataxia.[1] Vitamin B1 deficiency is responsible for this condition, and it develops usually on prolonged intravenous (IV) fluid therapy without vitamin supplementation. Although it is most commonly seen in chronic alcoholics and patients with gastrointestinal diseases, prolonged hyperemesis gravidarum is reported as a known cause for WE.[1] We report a case of WE presented with ocular manifestations that responded dramatically to thiamine replacement.

Case Report
A 29-year-old primigravida at 18 weeks of gestation was referred to ophthalmology clinic with altered sensorium, generalized weakness, difficulty in walking and blurring of vision for 15 days. She was admitted to a local hospital with excessive vomiting and hematemesis which was managed conservatively with IV fluids and antiemetics. On examination at arrival, she was not oriented to person and time, responded poorly to commands, and spontaneous speech was reduced. She was afebrile with a pulse rate of 126/min and blood pressure of 130/80 mmHg. Abdominal examination revealed a uterine size of 18 weeks gestation, and fetal heart was heard.

Ophthalmic examination showed a visual acuity of counting fingers at 1 meter in both eyes. There was no nystagmus; extraocular movements showed bilateral restriction of adduction. Pupillary reaction was normal. Fundus examination in right eye revealed mild blurring of the disc margin associated with peripapillary hemorrhages and exudates [Figure 1a]. Left eye showed marked blurring of disc margin along with peripapillary hemorrhages, exudates, and macular edema [Figure 1b]. Neurological examination revealed ataxic gait, muscle tone was decreased, and power was 4/5 for both upper and lower limbs. Sensory examination was normal and there were no involuntary movements.

Blood investigations revealed mild anemia and leukocytosis with low serum sodium and potassium levels. Urine dipstick showed 3+ ketones and microscopy showed...
18–20 WBC’s/hpf. Urine culture revealed no growth. Ultrasonogram confirmed a single live intrauterine fetus of 18 weeks size.

Nerve conduction study was normal. Magnetic resonance imaging (MRI) of the brain revealed hyperintensities in bilateral thalami and mamillary bodies on fluid-attenuated inversion recovery (FLAIR) and T2-weighted (T2W) images [Figure 2]. A diagnosis of WE with ophthalmic changes secondary to hyperemesis gravidarum was made.

She was started on injection thiamine 500 mg IV three times daily and switched over to Tablet Thiamine 100 mg three times after 5 days. Her condition began to improve after 3–4 days, optic disc edema decreased and hemorrhages reduced. Fundus photography 2 weeks after starting treatment revealed complete resolution of the fundus changes [Figure 3a and b]. She was discharged on oral thiamine 100 mg daily.

**Discussion**

WE was reported for the first time in 1881 by Carl Wernicke as an illness that consisted of paralysis of eye movements, ataxia, and mental confusion associated with Thiamine deficiency.[2] Thiamine deficiency can cause intra- and extra-cellular edema, glial cell proliferation, neuronal demyelination, and cellular degeneration. Most cases of WE are related to chronic alcoholism. However, it is crucial to recognize other rare causes such as malignancy, disseminated tuberculosis, acquired immunodeficiency syndrome, starvation, chronic hemodialysis, glucose infusion to a chronically malnourished person and persistent vomiting such as hyperemesis gravidarum.[3,4]

In the diagnosis of WE, the classic triad of symptoms-ocular abnormalities, confusion, and ataxia are reported in up to 66% of cases.[2] Additional neurological findings include decrease in deep-tendon reflexes, loss of tonus and dysarthria. Rarely, optic disc edema and retinal hemorrhages may be seen. Papillitis due to thiamine deficiency may lead to progressive visual loss and ultimately, optic atrophy. Occasionally, visual deterioration may be the first sign of thiamine deficiency.[2]

The ocular muscle and gaze palsies are attributed to lesions of sixth and third nerve nuclei whereas nystagmus is due to lesions in the region of vestibular nuclei. These lesions are characterized by lack of significant destruction of nerve cells, resulting in rapid recovery on thiamine replacement.[3]

MRI Brain is usually diagnostic and shows symmetrical hyperintensities in the dorsomedial nucleus of thalami, midbrain tectum, periaqueductal gray matter, around the fourth ventricle, and in the mamillary bodies in T2W and FLAIR images.[6,7]

Delay in the initiation of treatment could lead to irreversible neurological sequelae and visual impairment. This is especially important in pregnancy, as it could result in miscarriage, intrauterine growth retardation and fetal death.[3,4]
In conclusion, WE should be suspected in pregnant women with hyperemesis gravidarum if they present with neurological symptoms, visual loss, retinal hemorrhage or ocular muscle palsy and high-dose thiamine supplement must be started at the earliest to avoid sequelae.

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**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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