Case Report:
A Rare Case of Wernicke’s Encephalopathy Due to Hyperemesis Gravidarum.

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Citation

Abstract: A young pregnant woman with hyperemesis gravidarum presented to casualty with ataxia, confusion and diplopia. Examination revealed nystagmus and bilateral retinal haemorrhages. Characteristic brain MRI findings and rapid response to thiamine was suggestive of Wernicke’s encephalopathy due to hyperemesis gravidarum. Wernicke’s encephalopathy is a potentially reversible condition caused by thiamine deficiency. It is usually suspected in the setting of chronic alcoholism and might not be recognised when associated with other conditions. A high index of suspicion is required since lack or delay of treatment may lead to high morbidity and mortality.

Key Words: Wernicke’s Encephalopathy, Hyperemesis gravidarum, Thiamine deficiency.

Introduction: Hyperemesis gravidarum is associated with long lasting and uncontrollable nausea and vomiting with the common complications like dehydration, ketosis, hypochloremia and hypocalcemia which may cause an increase in maternal mortality and morbidity. Wernicke’s encephalopathy which manifests with diplopia, amnesia, confusion, apathy and ophthalmoplegia is one of the rarest complication of hyperemesis gravidarum due to thiamine deficiency. Wernicke’s encephalopathy is usually associated with chronic alcoholism. Other causes include malnutrition, starvation, malignant diseases, intravenous feeding and consumption of thiamine deficient diet. Wernicke’s encephalopathy due to severe Hyperemesis gravidarum is reported uncommonly with the incidence of 0.1 – 0.5%. [1]

We present here a case of Wernicke’s encephalopathy due to Hyperemesis gravidarum with intrauterine death who recovered without any neurological sequelae due to timely diagnosis and appropriate treatment.

Case Report:
A 29 years old housewife presented to us in the 17th week of her second pregnancy with the history of altered sensorium. Further history obtained revealed that she had unsteadiness of gait few days prior to admission. She was treated as a case of Hyperemesis gravidarum as she had been vomiting since 8th week of her pregnancy continuously despite various antiemetics. There was no history of fever or alcohol consumption.

On admission she was drowsy and unable to obey simple commands. Her BP recorded 118/76 mm of Hg. However she moved all four limbs on painful stimuli. There was neither meningism nor cerebellar signs. The reflexes were normal with negative Babinski’s response. Nystagmus was present in both horizontal and vertical directions but the range of eye movements were normal. Her BP recorded 118/76 mm of Hg. However she moved all four limbs on painful stimuli. There was neither meningism nor cerebellar signs. The reflexes were normal with negative Babinski’s response. Nystagmus was present in both horizontal and vertical directions but the range of eye movements were normal. Ultrasound abdomen done revealed intrauterine foetal demise which was delivered completely by obstetrician through induction. Based on clinical findings differential diagnosis of septic encephalitis and cerebral venous thrombosis were thought of.

CSF analysis was normal. MRI Brain plain revealed bilateral symmetrical T2 and FLAIR hyperintensities noted in medial thalami, tectum, mamillary bodies, hypothalamus and periaqueductal grey matter which was suggestive of Wernicke’s encephalopathy. MR Venography was normal. Other laboratory investigations were unremarkable.
In 1997, Caine et al proposed an operational criterion for the recognition and diagnosis of Wernicke’s encephalopathy accordingly; Wernicke’s encephalopathy is recognized if there are two of the following four signs; (i) dietary deficiencies, (ii) oculomotor abnormalities, (iii) cerebellar dysfunction, and (iv) either an altered mental state or mild memory impairment.[3]

Although most cases of Wernicke’s encephalopathy seen today are related to chronic alcoholism, it is vital to recognize other rare causes of this condition, such as systemic diseases (malignancy, disseminated tuberculosis, acquired immunodeficiency syndrome (AIDS)), starvation (anorexia nervosa, prisoners of war, schizophrenia, terminally ill cancer patients), iatrogenic (refeeding after starvation, chronic haemodialysis), and persistent emesis such as hyperemesis gravidarum. The prevalence of Wernicke’s encephalopathy in a non-alcoholic patient varies from 0.04% to 0.13%.[4]

Wernicke’s encephalopathy in a patient with hyperemesis gravidarum was first described by Sheehan in 1939.[5] To our knowledge, very few cases of Wernicke’s encephalopathy during pregnancy have so far been reported in the literature.[6]

The mechanism by which thiamine deficiency causes the focal neuropathy lesions found in Wernicke’s encephalopathy might be multiple.[7] Thiamine is an important co-enzyme for three critical enzymes in the Kreb’s and pentose phosphate cycle: transketolase, ketoglutarate dehydrogenase, and pyruvate dehydrogenase complex. Deficiency of thiamine and hence deficiency of these enzymes result in focal lactic acidosis, cerebral energy impairment, depolarization of neurons due to n-methyl- D-aspartate receptor mediated excitotoxicity. Ultimately, it results in alteration of blood brain barrier, generation of free radical, prompting cell death by necrosis and apoptosis.[7]

The body has approximately 18 days of thiamine storage. It is well understood that thiamine requirements are increased during pregnancy, and even more by the impaired absorption due to hyperemesis gravidarum.[8] Thiamine dependence is also increased in conditions with high metabolic rates and high glucose intake, and therefore its depletion due to reduced intake as well as IV dextrose administration results in thiamine deficiency and Wernicke’s encephalopathy.[9,10]

The treatment for Wernicke’s Encephalopathy includes high doses of thiamine, 300 mg per day, given eighth hourly for two days followed by 250 mg per day once daily until the patient tolerates oral thiamine.[12]

Our patient presented with the classical clinical triad following intractable vomiting and dextrose administration without thiamine supplementation. MRI imaging also detected sensitive neurological changes, raising the suspicion of an acute stage of thiamine deficiency. These findings were important for prompt diagnosis and treatment of our patient’s condition. Indeed there are reports of the usefulness of MRI imaging in diagnosing cases of Wernicke’s encephalopathy.[11]

Wernicke’s encephalopathy should be suspected in any nutritionally compromised patient who shows altered mental status before manifestation of the classical triad as, if left untreated, it could lead to ever irreversible and persistent neurological sequel or death. We would like to emphasize the importance of prompt thiamine supplementation in pregnant women with prolonged vomiting in pregnancy, especially before starting intravenous or parenteral nutrition. We would like to emphasize the importance of prompt thiamine supplementation in pregnant women with prolonged vomiting in pregnancy, especially before starting intravenous or parenteral nutrition.

In 1939, Sheehan described a syndrome of ‘Wernicke’s encephalitis hemorrhagica superioris’. [2]
References: