Chronic Alcoholism Induced Wernicke's Encephalopathy- A Case Report

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Abstract

Wernicke encephalopathy is an acute neurological condition caused due to thiamine deficiency. They can be characterized by a clinical trial of Ophthalmoplegia, ataxia and confusion. A 46years old male patient came with the complains of altered sensation, vomiting 2-3 episodes and loose stools in the past 2 days. He has been an alcoholic for the past 15 years. Systemic examination revealed drowsiness and irritability in patient and lab investigation revealed hyperammonaemia, abnormalities in liver and kidney function and traces of ketone bodies. In this case diagnosis of WE have been done with the help of blood tests and presented signs and symptoms. Treatment was initiated by discontinuing alcohol intake and administration of Inj. thiamine 500mg TID for a week and followed by T. thiamine 250mg along with other supportive treatment. This case report highlights the effect of thiamine deficiency and the critical role in the management thiamine deficiency, and if left untreated can lead to severe complication such a Korsakoff syndrome and death.

Keywords: *Wernicke encephalopathy, thiamine deficiency, alcoholism, Wernicke-Korsakoff syndrome*

1. Introduction

Wernicke's encephalopathy (WE) is a neuropsychiatric disorder caused due to thiamine deficiency which is characterized clinically by the triad of ocular abnormalities, ataxia and disturbances of consciousness. All patients with a history of alcoholism, malnutrition, malabsorption, tumour, inflammation, or other severe disease should be monitored for thiamine deficiency.[1]

Wernicke-Korsakoff syndrome is disorder that includes Wernicke encephalopathy which represents the "acute" phase and Korsakoff amnesic syndrome represents the disorder progressing to a "chronic" or long-lasting stage in distinct circumstances but various phases of the same illness (Wernicke-Korsakoff syndrome).[2]

2. Epidemiology

Data regarding, WE prevalence throughout the world are limited. However, postmortem histological analyses have provided evidence that the occurrence of WE are about 1% in the general population and 12.5–35.0% in alcohol-dependent patients. WE due to alcohol misuse is more common in males with a ratio of 1.69:1, whereas non-alcohol related causes are more common in females with a ratio of 1.84:1. Although the age of onset of WE differ with disease causation, the average age of onset of the condition is 40 years in alcohol-dependent patients, but in younger age groups, reasons unrelated to alcohol use are more prevalent. [8]

Studies also suggests that the prevalence of WE is mostly seen in Australia, Austria, Brazil, France, Germany, Norway and the United States with a prevalence from 0.6 to 2.2% based on autopsies.[4][8]

3. Etiology

WE is caused as a result of thiamine deficiency. Chronic alcoholism being the most commonest etiologic which accounts up to 90% factor associated with WE it can also occur in any patient with a nutritional deficiency state such as hyperemesis gravidarum, intestinal obstruction, and malignancy. bariatric surgery, starvation, anorexia nervosa, long term total parenteral nutrition, acute pancreatitis, Crohn's disease, thyrotoxicosis.[5][7]

4. Pathophysiology

Thiamine (vitamin B1) is a coenzyme that is essential for intricate organic pathways and plays a pivotal role in cerebral metabolism. The depletion of thiamine stores occurs 2-3 weeks after stopping the intake of thiamine, which leads to metabolic imbalances causing neurologic complications as a result of decreased levels of alpha-keto-glutarate, acetate, citrate, acetylcholine and accumulation of lactate and pyruvate, since This vitamin acts as a cofactor for several enzymes in the Krebs cycle and the pentose phosphate pathway, including alpha-keto-glutamic acid oxidation and pyruvate decarboxylation.[6]

Therefore, thiamine deficiency causes cytotoxic and vasogenic oedema which leads to bilaterally symmetrical lesions in the paraventricular regions of the thalamus, hypothalamus, mammillary bodies, periaqueductal region and floor of the fourth ventricle.[9]

4.1 Signs and symptoms

The recognition of WE is usually takes place at autopsy. The most common characteristic of this disorder is change in mental status. [7] The other symptoms include cognitive changes ranging from apathy and mild neurocognitive symptoms to severe symptoms including, in rare situations, coma. [8]

Ophthalmoplegia, is considered to be the second most common characteristics but other ocular findings may be present. Occurrence of complete ophthalmoplegia is rare, however horizontal nystagmus is the most common ocular abnormality.[8] The other ocular findings include sixth nerve palsy, ptosis, retinal haemorrhage, papilledema, anisocoria, or miosis. Characteristics such as gait ataxia, which ranges from mild gait abnormality to a complete inability to stand are considered to be the third or final characteristics.

In addition to above characteristics, presence of hypothermia, hypotension and coma can also be considered while suspecting the disease. [5] Approximately of 80% of patients who goes untreated with WE develop into korsakoff syndrome, characterized by memory impairment associated with confabulation. [5]

4.2 Diagnosis

Diagnosis of WE is considered to be difficult because the classical triad of signs (confusion, ataxia and ophthalmoplegia) occurs in only 10% of cases.[3]

Changes in mental status are the most commonly seen in both alcoholic and non-alcoholic WKS patients, and the changes include global confusion state (disorientation, apathy, drowsiness, indifference etc), memory disorder (mild memory impairment, amnesia), anxiety, fear, stupor. Ophthalmological abnormalities include nystagmus, lateral rectus palsy, conjugate gaze palsies, papillary abnormalities, retinal haemorrhage, ptosis, blurred vision, photophobia, diplopia and complete ophthalmoplegia. Other clinical symptoms that helps in the diagnosis of WE involve unsteadiness of gait, dysdiadochokinesis, impaired heel- shin testing etc, all resembles ataxia. [7]

Test for thiamine deficiency including whole blood or erythrocyte can be helpful in patients with suspected WE. MRI scan can be used to assess any neurological abnormalities by depicting the presence of lesions that results in brain damage and the presence of increased T2 signal (signifying oedema) can be seen at the paraventricular region of thalamus, hypothalamus and the periaqueductal region, while the cerebellum and mamillary bodies maybe in reduced size. However, it should also be taken into account that the presence of lesions due to brain damage depends person to person and are only seen in 58% of the patient. [8]Computed tomography (CT) is not much beneficial in detecting WE [7]

As a result, Caine proposed an operational criterion to help in the diagnosis of WE in chronic alcoholics. It is defined as patient who drank more than 80g of ethanol daily for most of their adult life, and the diagnosis of WE is based on two put of four signs which includes eye signs, cerebral signs, mild memory impairment or confusion and signs of malnutrition. [7]

5. Treatment

Since WE is accounted as a medical emergency and because of the rapid progression of disease, it is recommended that therapeutic administration of thiamine be commenced in any case where thiamine deficiency is suspected, even before a firm diagnosis has been made. Parenteral delivery is required because individuals may not have the best mechanisms in place to absorb the vitamin through the GI tract. There is no particular dosing or duration of vitamin B_1 to treat WE, however those with WE caused by alcohol use may need higher daily doses. The British authors have recommended 500 mg of thiamine three times a day for two to three days, followed by 250 mg daily until improvements stop, while the European Federation of Neurological Societies recommends intravenous administration of 200 mg of thiamine three times a day until there are no additional improvements in clinical conditions.[8]

6. Case report

A 46 year old male patient was presented with complains of altered sensation, vomiting 2-3 episodes and loose stools since past two days. His personal history revealed that he has been an alcoholic for the past 15 years.

The patient was reviewed in ICU, where he was disoriented/drowsy and irritable with limbs responding to painful stimulations, maintaining saturation in room air with SPO2 94%. His physical examination revealed pallor and icterus. General examination depicted a BP of 130/80 mmHg, Pulse rate 111 bpm and respiratory rate 22 cpm. Systemic examination revealed CNS- E_1 V_3 M_3 on Glasgow coma scale with CVS- S_1 S_2 heard and respiratory system showing positive NVBS.

Table 1: Biochemistry

Investigations	Results	Reference range
ammonia	77 µmol/l	9 - 30
urea- serum	76 mg/dl	12.9 - 42.8
creatinine- serum	1.9 mg/dl	0.7 - 1.2
total bilirubin	6.4 mg/dl	0.3 - 1.3
direct bilirubin	5.1mg/dl	0.1 - 0.4
indirect bilirubin	1.3 mg/dl	0.2 - 0.9
ast/sgot	885 u/l	upto 35
alt/sgpt	549 u/l	upto 45
alkaline phosphate	142 u/l	53 -128
ggt	551 u/l	<55
grbs	280 mg/dl	70-100
serum- sodium	149 meq/l	136 - 145
chloride- serum	117 meq/l	98 - 107
triglycerides	392 mg/dl	<150
vldl	78.4 mg/dl	<30
crp test quatitative	18.8 mg/l	<5
lipase- serum	1174 u/l	23 - 300
amylase- serum	118 u/l	31 - 107

An abdomen and pelvis ultrasound revealed Cystitis with Grade 1 fatty liver. Laboratory investigation revealed abnormalities in Haematology, Biochemistry and urine routine as shown above

Table 2: Haematology

CBC	Results	Reference range
platelet count	121 x10^3µl	150-450
neutrophil	81%	40-75
lymphocytes	14%	20-45

Table 3: Urine routine

investigations	results	reference range
colour	dark yellow	pale yellow
clarity	turbid	clear
protein	present (++)	neg
ketone bodies	traces	neg
leucocytes	10 – 15/ hpf	0-2
rbc	plenty	0 – 1/ hpf

He was diagnosed with Wernicke encephalopathy with the help of subjective and objective evidence. His treatment was initiated with Inj. Pantoprazole 40mg OD and Inj. Thiamine 500MG TID for a week, which was then changed to Tablet thiamine RH- ones a day. Inj. Ceftriaxone 2g BD and tablet Rifagut 550mg BD was given for 3 days followed by Inj. Piptaz 4.5g initially for 3 days then tapered to 2.25g for the next two days. He was also administered with tablet Udihep 300mg BD and Inj. Hepamerz TID as a hepato-protectant.

The patient complained of multiple thick yellowish foul-smelling crusts and plaques on the scalp which was diagnosed to be Pityriasis amiantacea and prescribed with Fucidin cream BD for 5 days. He also complained of constipation which was induced by Inj. Pantoprazole and was prescribed with syrup looz. Upon discharge he was prescribed with tablet Acamprostate 333mg and syrup Elcarno forte for 10 days. The patient showed improvement following treatment although he had positive cognitive deficit for which family members were counselled.

7. Discussion

A thiamine deficit can result in an acute neurological condition known as Wernicke's encephalopathy (WE). WE is characterised by a classical triad of symptoms such as altered mental state, ophthalmoplegia/nystagmus and ataxia/gait disturbances. According to Sara kohnke et. Al, although WE were described in the alcohol-dependent population, other groups of patients with no history of alcohol dependence, can also suffer from the condition such as conditions affecting nutrition, hyperemesis gravidarum and following bariatric surgery can also suffer from WE. Other causes include malignancies, immunodeficiency syndromes, liver diseases, prolonged parenteral nutrition, hyperthyroidism and severe anorexia nervosa.

The diagnosis of WE is clinically done on the basis of criteria proposed by Caine et.al in 1997 which requires at least two of the following features to be eligible for diagnosis, this includes dietary deficiency, eye signs, cerebellar signs and mild memory impairment or altered mental state. In this case the patient was presented with thiamine deficiency, altered mental state and cerebellar signs.

Treatment of WE should be commenced parenterally at any state where thiamine deficiency is suspected at higher daily doses until clinical improvements are ceased, followed by oral thiamine 500mg for 2-3days followed by 250mg per day. Administration of tablet Acamprosate 333mg has also shown significant improvement in patient's cognitive function and overall well-being.

8. Conclusion

We are being a medical emergency should be treated with high doses of IV thiamine as soon as thiamine deficiency is suspected since it is challenging to diagnose WE and necessitates combining clinical symptoms, MRI findings, and biochemical data. WE when left untreated can lead to a chronic condition called as Wernicke-Korsakoff syndrome in 80% of the patients which is characterized by memory impairment associated with confabulation and significant deficits in anterograde and retrograde memory. On the other hand, survivors of WE could experience Korsakoff psychosis and need ongoing clinical attention. Among them, less than 10% will benefit from long-term care.

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