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Korsakoff's Syndrome: A Clinical Diagnosis

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Abstract

In this review, we present an inpatient survey on Korsakoff's Syndrome (KS), an active and potentially life-threatening presentation in emergency department of a neurological tertiary care center. We describe the history of KS and its definition, its epidemiology, and a lack of consensus criteria for its diagnosis. The cognitive and behavioral symptoms of KS, which include anterograde and retrograde amnesia, executive dysfunction, confabulation, apathy as well as affective and social cognitive impairments are discussed. We present supportive radiological evidence and pharmacological treatment protocol we followed for the patient. Our review shows that thiamine deficiency is a crucial factor in etiology of KS. Although alcohol abuse has certainly buttressed the condition. In this review we compared day wise progression to good health after starting proper treatment.

Keywords: ***Abbreviated Mental Test (AMT); Alcohol use disorder; Korsakoff's syndrome; Mammillary bodies atrophy; MRI findings for Korsakoff syndrome; Thiamine deficiency; Wernicke encephalopathy***

1. Case Summary

A 63-year-old male from Palpa district, Nepal presented to emergency department of a neurological center with complains of being quiet and withdrawn, lethargic and disturbed orientation to self and surroundings since yesterday. At the time of presentation in emergency, he was restless and agitated. He was unable to recognize his family and surrounding where he was brought to. He was unaware of the long-distance travel that he had already made to reach to our center. His youngest son explained this has been happening since yesterday. They visited a medical center in Palpa district which referred this to our center with a working diagnosis of alcohol use disorder. He (son) explains he is disoriented and has been having disturbed sleep awake cycle. He consumes local alcohol daily and in a huge amount. He had similar episode 5 months back for which he was treated symptomatically in the medical center and was advised to stop taking more alcohol. The consequences were explained. But the patient continued taking alcohol despite the warnings and advice that were given. Patient explains it has been more

than 40 years since when he started taking alcohol and has been constantly increasing his amount of intake. He is a known diabetic and hypertensive patient for which he has been taking antidiabetic (Tab. Metformin 500 mg OD) and anti-hypertensive (Tab Amlodipine + Losartan 5/50 mg OD).

Patient was lethargic hence all examinations could not be accessed at time of presentation in emergency department. On Glasgow Coma Scale E2V4M6. On head-to-toe examination horizontal nystagmus was present however no signs of gaze palsy or ophthalmoplegia. During the survey there were no significant positive finding of self-harm or injury due to fall. No signs drug abuse.

His cerebellar signs showed ataxic gait, abnormalities in past pointing, dysdiadokinesia and impaired hell shin testing.

To check his cognitive status Abbreviated Mental Test (AMT) was undertaken (2/10).

TABLE 1. **AMT criteria at time of admission.**

Sn.	AMT categories	Yes/No
1	He could say his age	Yes
2	Time of day	No
3	Year	No
4	Place	No
5	Recognition of people	No
6	Date of birth	Yes
7	National day	No
8	Prime minister	Yes
9	Counting backwards from 20 to 1	No
10	Recall of address	No

Modified Oxford Handicap Scale (MOHS) of 3 during the episode.

Regular baseline investigation along with radiological investigation were done, medications started and was admitted to intensive care unit.

The list of medication used during the hospital stay were:

1. Inj. THIAMINE (B1) 1 ampule in each drip of NS x 3 drip per day for 2 days
2. Tab. HALOPERIDOL 0.25 mg twice daily for 5 days
3. Tab. THIAMINE 100 mg three times a day for 30 days
4. Tab. CLOBAZAM 5 mg at night before sleep for 30 days

All the examinations were repeated on 3rd day of admission. A lot of findings were improved as mentioned below. On Glasgow Coma Scale E4V4M6. On head-to-toe examination horizontal nystagmus was present however no signs of gaze palsy or ophthalmoplegia. During the survey there were no significant positive finding.

His cerebellar signs showed slight improvement in ataxic gait, abnormalities in past pointing, dysdiadokinesia and impaired hell shin testing.

To check his cognitive status Abbreviated Mental Test (AMT) was undertaken (8/10).

TABLE 2. **AMT criteria on 3rd day of admission.**

Sn.	AMT categories	Yes/No
1	He could say his age	Yes
2	Time of day	Yes
3	Year	Yes
4	Place	Yes
5	Recognition of people	Yes
6	Date of birth	Yes
7	National day	Yes
8	Prime minister	Yes
9	Counting backwards from 20 to 1	No
10	Recall of address	No

On the 6th day of admission, he was discharged and asked to follow up in OPD in 1 month for further evaluation.

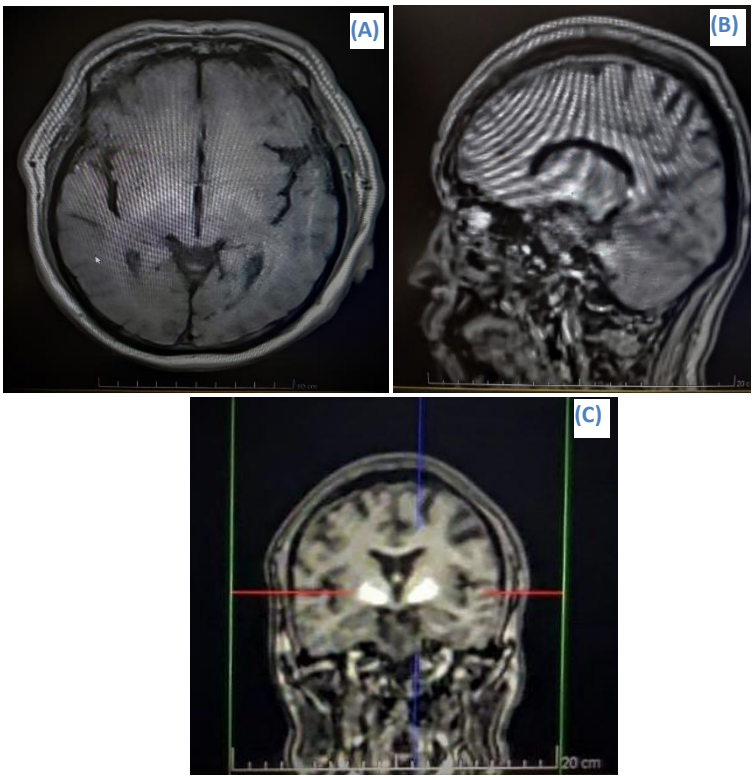

FIG. 1. **(A) This is the T1 (A) Axial Flair MRI Image of Patient Which Shows Symmetrically Increased Intensity Over The Mamillary Body Region In (B) Sagittal (C) Coronal.**

2. Introduction

The residual syndrome in the patient who suffered from Wernicke Encephalopathy (WE) but didn't receive immediate and adequate treatment with thiamine replacement therapy is Korsakoff's Syndrome (KS). The KS symptoms usually presents with retrograde and antegrade amnesia along with cognitive and behavioral deficits which severely affects day to day activities. Benon R. and LeHuché R [1] described the characteristic signs of Korsakoff syndrome with some additional features including: confabulation (false memories), fixation amnesia, paragnosia or false recognition of places, mental excitation, and euphoria.

Thiamine is essential for the decarboxylation of pyruvate, and deficiency during this metabolic process is thought to cause damage to the medial thalamus and mammillary bodies of the posterior hypothalamus, as well as generalized cerebral atrophy. These brain regions are all parts of the limbic system, which is heavily involved in emotion and memory. KS involves neuronal loss, that is, damage to neurons (gliosis) which is a result of damage to supporting cells of the central nervous system and hemorrhage or bleeding also occurs in mammillary bodies. Damage to the medial dorsal nucleus or anterior nuclei of the thalamus (limbic-specific nuclei) is also associated with this disorder. Cortical dysfunction may have arisen from thiamine deficiency, alcohol neurotoxicity, and/or structural damage in the diencephalon [2,3].

KS causes deficits in declarative memory in most people, but leaves implicit spatial, verbal, and procedural memory functioning intact [4]. People with KS have deficits in the processing of contextual information. Context memories refers to the where and when of experiences and is an essential part of recollection. The ability to store and retrieve this information, such as spatial location or temporal order information, is impaired. Research has also suggested that people with Korsakoff syndrome have impaired executive functions, which can lead to behavioral problems and interfere with daily activities. It is unclear, however, which executive functions are affected most.

At first it was thought that those with KS used confabulation to fill in memory gaps. However, it has been found that confabulation and amnesia do not necessarily co-occur. Studies have shown that there is dissociation between provoked confabulation, spontaneous confabulation (which is unprovoked), and false memories [3]. That is, people affected could be led to believe certain things had happened which actually had not, but so could people without Korsakoff syndrome.

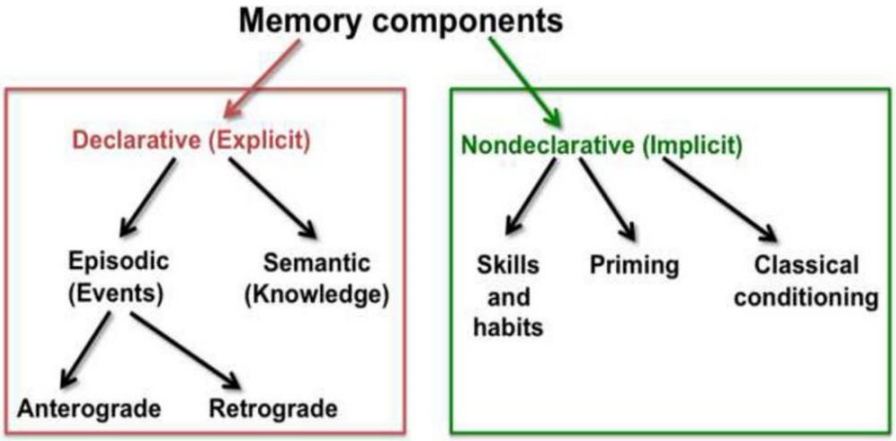

FIG. 2. **Component processes of memory. In ks, declarative (explicit) memory components (in the red box) are impaired while non declarative (implicit) memory components (in the green box) are relatively spared. This review focuses on anterograde episodic processes [5].**

TABLE 3. **The operational criteria for the clinical diagnosis of WE, as formulated by Caine et al [6].**

Symptom or sign	As evidenced by one or more of the following
Dietary deficiencies	– Undernutrition (body mass index <2 SD below normal)
	– A history of grossly impaired dietary intake
	– An abnormal thiamine status
Oculomotor abnormalities	– Ophthalmoplegia
	– Nystagmus
	– Gaze palsy
Cerebellar dysfunction	– Unsteadiness or ataxia
	– Abnormalities of past pointing
	– Dysdiadokokinesia
	– Impaired heel-shin testing
Either an altered mental state	– Disorientation in two of three fields
	– Confused
	– An abnormal digit span
	– Comatose
Or	Or
Mild memory impairment	– Failure to remember two or more words in the four-item memory test
	– Impairment on more elaborate neuropsychological tests of memory function

Notes : When two out of these four criteria apply, the clinical diagnosis of WE is made. The criteria are less sensitive in case of a co-occurring hepatic encephalopathy. **Abbreviation :** WE, Wernicke encephalopathy.

3. Causes

Conditions resulting in thiamine deficiency and its effects include chronic alcoholism and severe malnutrition. Alcoholism may co-occur with poor nutrition, which in addition to inflammation of the stomach lining, causes thiamine deficiency. Other causes include dietary deficiencies, prolonged vomiting, eating disorders, and the effects of chemotherapy. It can also occur in pregnant women who have a form of extreme morning sickness known as hyperemesis gravidarum. Mercury poisoning can also lead to Korsakoff syndrome. Though it does not always co-occur, this disorder can emerge frequently as a consequential result of Wernicke's encephalopathy.

A number of factors may increase a person's risk to develop Korsakoff syndrome which includes age, alcoholism, chemotherapy, dialysis, extreme dieting, genetic factors. These factors are often related to general health and diet.

PET scans show that there is a decrease of glucose metabolism in the frontal, parietal and cingulated regions of the brain in those with Korsakoff syndrome. This may contribute to memory loss and amnesia. Structural neuroimaging has also shown the presence of midline diencephalic lesions and cortical atrophy [2]. MRI shows bilateral increased intensity in the mamillary bodies, dorsal medial thalamus, and periaqueductal region.

Structural lesions of the central nervous system, though rare, can also contribute to symptoms of KS. Severe damage to the medial dorsal nucleus inevitably results in memory deficit. Additionally, autopsies of people who had KS have showed lesions in both the midline and anterior thalamus, and thalamic infarctions. Bilateral infarctions to the thalamus can result in Korsakoff-induced amnesia as well. These findings imply damage to anterior thalamic nuclei can result in disruptive memory.

4. Treatment

Basically, the treatment is based on supplement of thiamine and stopping of alcohol intake. Proper counselling regarding the behavioral changes and medication must be given. Periodic psychiatric evaluation must be taken in account.

5. Learning Point

- Korsakoff Syndrome is acute and life-threatening neurological condition.
- Operational criteria and Abbreviated Mental Test for clinical diagnosis.
- Thiamine deficiency being a crucial factor for symptoms.
- Radiological evidence supporting Korsakoff Syndrome.
- Management protocol in tertiary neurological hospital of Nepal.

6. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patient 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.

7. Financial Support and Sponsorship

No fund was needed.

8. Conflict of Interest

There are no conflict of interest.

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