(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 5, Issue 6 , Page No: 484-486
November-December 2022

IJMSCR

Cadasil In Alcohol Dependence Mimick Wernicke's Encephalopathy

[1]Dr. Seytha Najva, [2]Dr. Arun Seetharaman, [3]Dr. Sridhar, [4]Dr. Agila C.

***Corresponding Author:**
Dr. Arun Seetharaman

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

CADASIL - Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy, a rare genetic condition with autosomal dominant inheritance characterised by migraine with aura, urinary incontinence, repeated seizures, recurrent stroke, pseudo bulbar palsy, and dementia with gait disturbances. Wernicke's encephalopathy is neurological condition encountered in chronic alcohol dependence case characterized by ataxia, ophthalmoplegia, and global confusion. In this case report, we highlight a 44years old male, known case of alcohol dependence syndrome presented with behavioural changes, sialorrhea, memory disturbances and slowness of movements with psychotic features. Patient was initially diagnosed as a case of Wernicke's encephalopathy and Alcohol induced psychosis, however MRI- brain revealed a picture of CADASIL, which was confirmed by further investigation. The implication of this case report is that alcohol dependence syndrome can be presented with multiple comorbidities, hence investigating and ruling out organicity in any substance abuse and dependence/ psychiatric disorder helpful to prevent further deficits and early treatment options.

Keywords: CADASIL, leukoencephalopathy, subcortical infarcts

Introduction

CADASIL is a rare genetic condition with autosomal dominant inheritance. It is mainly caused due to mutation of NOTCH-3 gene which is located on chromosome 19q12. Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy is characterised by migraine with aura, urinary incontinence, repeated seizures, recurrent stroke, pseudo bulbar palsy, and dementia with gait disturbances. White matter changes in basal ganglia and subcortical white matter is detected by neuroimaging techniques. CADASIL may also result in personality and mood changes. Behavioural changes are also seen in 30% of cases with adjustment disorder, and mood disorders more common among depression. However CADASIL with psychotic features is known to be a rare combination which has been reported only in several articles. Disturbances in cortical and subcortical network due to multiple white matter infarcts can be given as an explanation for emergence of psychotic features and changes in reality monitoring.

Case Report

A 44/M with alcohol use for past 10yrs with dependence pattern for the past 2yrs, came with complaints of suspiciousness that wife is having extramarital affair, irritable & violent under alcohol influence for past 2yrs, difficulty in recalling things & confusion for pats 4wks. His last intake of alcohol was 4weeks back prior to admission with history of hearing voices & irrelevant speech for past 2 days. He had past history of multiple admission in private hospital, & de-addiction centre, relapse & poor compliance to drugs. Pre-morbidly he was an impulsive, irresponsible, sensitive person with suspiciousness, poor moral values, combativeness, and tendency to blame others. His general physical examination revealed BP-140/100mmHg; PR-100/min. Systemic examination revealed; CNS-disoriented, slowness of gait, mild abnormality in

finger- nose test, & dysdiadokinesia present. Mental Status Examination revealed increased psychomotor activity; SPEECH: increase in tone, tempo volume and decreased reaction time, AFFECT: lability present and restricted, THOUGHT: delusion of infidelity, PERCEPTION- Auditory & visual Hallucination present, COGNITION: attention not aroused/concentration not sustained, immediate memory & recent memory – mildly impaired with MMSE- 17/30. Initailay working diagnosis was Alcohol induced amnestic syndrome (werenicke's encephalopathy), Alcohol induced psychotic disorder. Inj.Thiamine 300mg TDS was given for 1 week & switched to oral 200mg TDS, patient's memory improved, but continued to have ataxia, and slowness of movements. Patient was treated with T.Risperidone 2mg & lorazepam. On 8th day - mild rigidity was present hence he was started on trihexyphenidyl 2mg. Patient recovered with no psychotic symptoms, memory was intact & hence discharged on 13th day with Thiamine 100mg TDS, T.Risperidone 2mg, Acamprosate 333mg 2 TDS. After 1 week on follow-up there was worsening of symptoms, immediate & recent memory was impaired, patient was disoriented with excessive salivation, crying spells, slowness of movements, decline in self-care, social and occupational functioning. Following which MRI was done which revealed foci of T2W/FLAIR hyperintensities involving cortical & subcortical regions of bilateral frontal, temporal & deep white matter regions (possibly CADASIL).

Discussion

400 families have been affected by CADASIL worldwide. It is caused due to the mutation of NOTCH3 gene. NOTCH3 gene plays an important role in the preservation of phenotype stability of vascular smooth muscle.[1] CADASIL is linked to more than 200 different mutations in NOTCH3 gene, especially the cysteine residue variant impairs tertiary protein structure and functions.[2] This neurological condition is associated with Ischemic stroke/ TIA, cognitive impairment, vascular dementia.[2] neurological studies such as MRI shows T2 hyperintensities in periventricular & subcortical white matter, & anterior temporal lobe[3]. Granular osmophilic deposits around blood vessels in a skin biopsy is a unique feature of CADASIL. These osmophilic deposits play an important role in sequestration of proteins responsible for blood vessel hemostasis.[4] CADASIL may also result in personality & mood changes. Behavioural changes includes 30% with adjustment & mood disorders, especially depression. CADASIL with psychotic features are rare. However associated psychotic features and changes in reality monitoring is mainly due to the disturbances in cortical & subcortical network due to multiple white matter infarcts,[1] which has been seen in this case. Schizophrenia and CADASIL has been linked by disruption of Notch-dependent transcription, which is important for many biological functions, suggesting a shared dysfunctional pathways.[5] Therapeutic implication of triptans & ergot-based drugs is due to potential possibility of vasoconstriction and higher risk stoke.[2]

Conclusion:

CADASIL is rare and distressing genetic condition without any specific treatment option except symptomatic approach. However ongoing translational research in genetics and advances in basic science will help to empower with knowledge on its complete pathophysiology, confirmatory specific biomarkers and therapeutic options for patients. The implication of this case report is that alcohol dependence syndrome can be presented with multiple comorbidities, hence ruling out organicity in any substance abuse and dependence/ psychiatric disorder to prevent further deficits and early treatment.

References

1. Mishra DK, Kishore A, Niranjan V. CADASIL syndrome (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy) presenting as psychosis. General psychiatry. 2018;31(3).
2. Zhu S, Nahas SJ. CADASIL: imaging characteristics and clinical correlation. Current pain and headache reports. 2016 Oct;20(10):1-5.
3. Wang MM. Cadasil. Handbook of clinical neurology. 2018 Jan 1;148:733-43.
4. Ferrante EA, Cudrici CD, Boehm M. CADASIL: new advances in basic science and clinical perspectives. Current opinion in hematology. 2019 May;26(3):193.

Volume 5, Issue 6; November-December 2022; Page No 484-486

5. Ho CS, Mondry A. CADASIL presenting as schizophreniform organic psychosis. General hospital psychiatry. 2015 May 1;37(3):273-e11

Volume 5, Issue 6; November-December 2022; Page No 484-486