https://doi.org/10.48047/AFJBS.6.14.2024.10575-10579

African Journal of Biological Sciences

Journal homepage: http://www.afjbs.com

Research Paper Open Access

CLINICAL SIGNIFICANCES AND PSYCHIATRIC MANIFESTATIONS IN A MIDDLE AGED PATIENT WITH CREUTZFELD JACOB DISEASE PRESENTING AS CATATONIA – A CASE REPORT

Gauri Gupta[1], Poonam Bharti[2]*, Vikas Kumar Pandey[3], Manish Kumar Maity[3], Aman Bharti[4]

[1]Junior Resident, Department of Psychiatry, MM Institute of Medical Sciences and Research, Maharishi Markandeshwar (Deemed to be University), Mullana - 133207, Ambala, Haryana, India

[2]Head of the Department & Professor, Department of Psychiatry, MM Institute of Medical Sciences and Research, Maharishi Markandeshwar (Deemed to be University), Mullana - 133207, Ambala, Haryana, India

[3]Research Scholar, Department of Pharmacy Practice, MM College of Pharmacy, MaharishiMarkandeshwar (Deemed to be university), Mullana - 133207, Ambala, Haryana, India

[4]Associate Professor, Department of Medicine, Guru Gobind Singh Medical College and Hospital, Faridkot - 151203, Punjab, India

***Address for correspondence:**
Dr. Poonam Bharti
Head of the Department & Professor,
Department of Psychiatry,
MM Institute of Medical Sciences and Research,
Maharishi Markandeshwar (Deemed to beUniversity),
Mullana - 133207, Ambala, Haryana, India
Email id – poonambharti109@gmail.com

Received: 15 June 2024 **Accepted**: 25 July 2024 **Published**: 29 Aug 2024

Volume 6, Issue 14, Aug 2024
doi: 10.48047/AFJBS.6.14.2024.10575-10579

ABSTRACT

Creutzfeld jacob disease (CJD) is a prion, neuro-degenerative disorder. This disease is rapidly progressive and always fatal. Infection with this disease leads to death usually within 1 year of onset of illness.

Here, we present a case report of a 50 years old female patient, who came to the psychiatry outpatient department with the complaints of poor self-hygiene, psychomotor retardation and rigidity in all limbs. Her differential diagnosis was kept as catatonic schizophrenia and meningitis; which was later confirmed as Creutzfeld jacob disease (CJD) by a detailed CSF analysis and

neurological examination.
Though CJD is not a common disease, it should be considered as differential diagnosis whenever neuropsychological manifestations, especially progressive decline in cognition, along with symptoms such as psychomotor retardation and hallucinatory behaviour are present.

Keywords - Cruetzfeld Jacob Disease, prion's disease,psychiatric manifestations.

INTRODUCTION

Creutzfeldt Jakob disease (CJD) is a rare, neurodegenerative, prion disease with an incidence of one person per million individuals in every year and mortality rate is 100%. It occurs mostly in sporadic form (90%) followed by familial (15%) and acquired forms (5%). Clinical onset is characterised in most cases by neurological symptoms, whereas in a much smaller percentage by signs of mental deterioration and psychiatric symptoms [1].

A diagnosis of CJD should not be neglected in elderly patients presenting with recent onset and rapid progression of behavioural changes, anxiety, irritability, mood deflection, and insomnia with no psychopathological history [2].

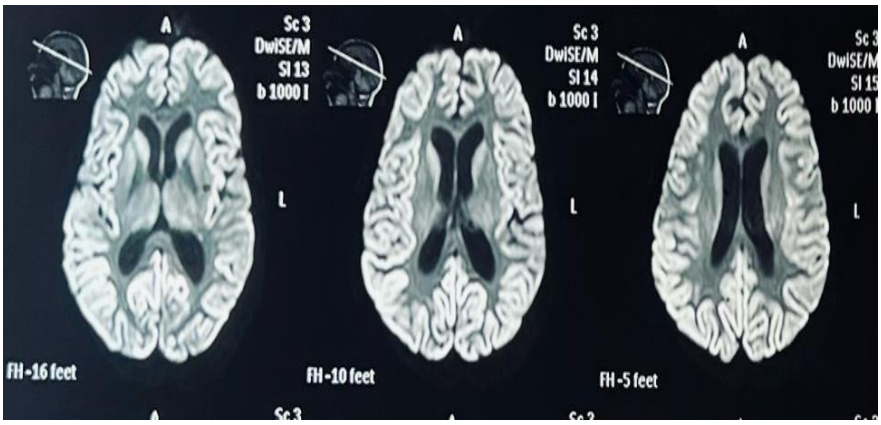
CASE PRESENTATION

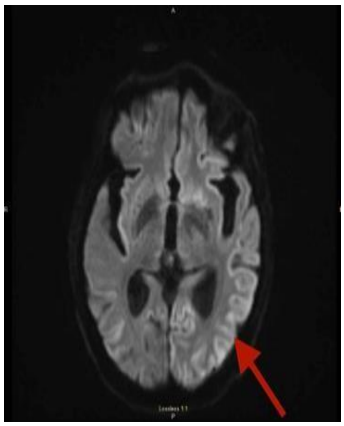
A 50 years old married, Muslim woman, was brought to the psychiatry outpatient department with history of disturbed self-hygiene, psychomotor retardation and rigidity. The patient has an 8 months old history of gradual onset of forgetfulness and confusion.

The patient also developed throbbing headache around a week before the date of hospitalization. Following which a sharp decline in communication was noted along with increased rigidity in limbs as she was not able to stretch her limbs fully which led to the patient being confined to her bed unable to carry out any household functions. She was no longer able to communicate properly other than occasional screams whenever she needed any assistance in basic biological functions such as eating food or visiting the washroom. The family history and past history for fever were unremarkable; however, the history of beef consumption was noted.

At first psychiatric evaluation, the patient was conscious and alert but not responding adequately for a detailed examination or cognitive functions to be carried out. On the Bush Francis Catatonia Rating Scale (a valid, reliable, and user-friendly catatonia rating scale), the total score was 25 with the patient scoring on immobility, staring, rigidity, combativeness. A detailed neurological evaluation was also done which revealed increased muscle tone in all four limbs with brisk reflexes. Bilateral plantar reflex for the patient was flexor along with a GCS score of E4M5V2. Due to quick progression of symptoms and abnormal neurological findings, the differential diagnosis of meningitis and catatonia were kept.

Meningitis was ruled out due to absence of any febrile episode and negative findings on elicitation of Kernig's as well as Brudzinski's signs. During the course of hospital stay, routine blood investigations, MRI with and without contrast were carried out along with CBC, LFT, RFT, Thyroid function tests, serum vitamins B12 levels which came out to be within normal limits. Ribboning was seen in the cortical region on MRI along with T2 weighted bilateral cortical hyper-intensities. During previous hospitalisations of the patient in other tertiary care centre, MRI contrast revealed increased diffusion signal in parieto-occipital regions which on comparison to the current CEMRI films was seen progressing to the frontal region as well. Brain MRI examination showing cortical ribboning pattern suggestive of CJD-CSF analysis for 14-3-3 protein was also carried out previously which came out to be positive along with periodic positive sharp wave with diffuse slowing on EEG, hence proving the diagnosis of CJD.





She was started on Quetiapine 25 mg in the morning and 50 mg at night along with a combination of Donepezil 5 mg and Memantine 10 mg.The patient was discharged on request due to lack of an adequate attendant to stay with the patient during hospital stay. She was discharged with an adequate management of sleep disturbances, irritability along with management of bed sores and adequate psychoeducation of her family members regarding maintenance of her hygiene.

DISCUSSION

Initially described in 1921, Creutzfeldt-Jakob disease (CJD) is a rare, transmissible prion disease of the brain [3]. The unusual syndrome of sporadic CJD (SCJD) is characterized by a rapidly progressive dementia, often accompanied by myoclonus and other signs of central nervous system (CNS) dysfunction, ultimately leading to death. Creutzfeldt-Jakob disease and its various forms have received increased media attention due to concerns regarding bovine spongiform encephalopathy (BSE) or mad cow disease, which has been found in U.S. and Canadian cattle populations [4,5].

The diagnosis of SCJD is based on clinical symptoms along with laboratory tests including EEG, imaging, and CSF analysis for 14-3-3 protein. On T2-weighted MRI, hyper intense areas could be found in the caudate nucleus, putamen, globus pallidus, and thalamus. Diffusion weighted MRI may indicate bilateral symmetrical hyper intense signals in the basal ganglia or cortex cerebri [6].

Wall et al. retrospectively reviewed 126 patients of sporadic CJD in Mayo Clinic from 1976 to 2001, and found that 26% of the patients had psychiatric symptoms at presentation while 80% demonstrated some psychiatric dysfunction within 100 days of onset of illness. Prominent psychiatric symptoms were sleep disturbances, psychosis, and depression [2].

Mehndiratta et al. reported 10 cases of CJD from North India where abnormal behaviour was seen in 70% cases [7].

Satishchandra et al. in their epidemiological study recorded 20 definite and 10 probable cases of CJD from 1971 to 1990. In that paper, the authors found that psychiatric manifestations were present at onset in 3 cases with 10 patients had visual complaints during the time of diagnosis. Clustering of cases was seen in Mumbai and Bengaluru which was more apparent than real [8].

In a study by Yen CF et al., four cases out of five presenting as CJD were sent to the psychiatric unit and were found to have symptoms of mood, thought, behaviour and perception during the course of their illness [9].

Shekhawat et al., have reported of a case of CJD presentation with catatonic signs in a North Indian patient. It is most likely, the psychiatrist who shall first encounter a patient presenting with catatonic symptoms who is eventually diagnosed as CJD [10].

CONCLUSION

In this case, the diagnosis was consolidated after neuroimaging and detailed neurological examination as well as CSF analysis findings. The patient was managed symptomatically for irritability, sleep disturbances and self-hygiene. In clinical practice, CJD should not be neglected as a differential diagnosis in adult and elderly patients who are referred to psychiatrists as catatonic symptoms maybe the presenting feature of the disease.

Conflicts of interest - There are no conflicts of interest

ABBREVATIONS

CJD	Creutzfeld Jacob Disease
CSF	Cerebro Spinal Fluid
CBC	Complete Blood Count
LFT	Liver Function Test
RFT	Renal Function Test
BSE	Bovine Spongiform Encephalopathy

REFERENCES

1. Jakob A. Uber eine der multiplen Sklerose klinisch nachestehende Erkrankung des Zentralnervensystems (spastische Pseudosklerose) mit bemerkenswertem anatomischen Befund. Medizinische Klinik. 1921;13:372-6.

2. Wall CA, Rummans TA, Aksamit AJ, Krahn LE, Pankratz VS. Psychiatric manifestations of Creutzfeldt-Jakob disease: a 25-year analysis. The Journal of neuropsychiatry and clinical neurosciences. 2005 Nov;17(4):489-95.

3. CB T. How now, mad cow? Time [Internet]. 2024 [cited 2024 Aug 31];163(2).Available from: https://pubmed.ncbi.nlm.nih.gov/14735655/

4. Donnelly CA. Bovine Spongiform Encephalopathy in the United States — An Epidemiologist's View. New England Journal of Medicine. 2004 Feb 5;350(6):539– 42.

5. Bruce ME, Will RG, Ironside JW, McConnell I, Drummond D, Suttie A, McCardle L,Chree A, Hope J, Birkett C, Cousens S. Transmissions to mice indicate that 'new variant'CJD is caused by the BSE agent. Nature. 1997 Oct 2;389(6650):498-501.

6. Weihl CC, Roos RP. Creutzfeldt–Jakob disease, new variant Creutzfeldt–Jakob disease, and bovine spongiform encephalopathy. Neurologic clinics. 1999 Nov 1;17(4):835-59.

7. Mehndiratta MM, Bajaj BK, Gupta M, Anand R, Tatke M, Seryam S, Nehru R, Puri V, Khwaja GA. Creutzfeldt-Jakob disease: report of 10 cases from North India. Neurology India. 2001 Oct 1;49(4):338.

8. Satishchandra P, Shankar SK. Creutzfeldt-Jakob disease in India (1971–1990). Neuroepidemiology. 1991 Apr 12;10(1):27-32.

9. Yen CF, Lin RT, Liu CK, et al: The psychiatric manifestation of Creutzfeldt- Jakob disease. Kaohsiung J Med SCI 1997;13:263-267.

10. Shekhawat LS, Kumar S, Ramdurg SI, Manjunatha N, Gupta P. New variant Creutzfeldt-Jacob Disease presenting with catatonia: a rare presentation. The Journal o Neuropsychiatry and Clinical Neurosciences. 2012 Jul;24(3):E51-3.