Journal of Dementia

Gonda, J Dement 2023, 7:5

Editorial Open Access

Creutzfeldt - Jakob Disease from Prions to Neurodegeneration

Kaokaf Gonda*

Department of Psychiatry and Biobehavioral Science, UCLA School of Medicine, USA

Abstract

Dementia is a progressive neurodegenerative disorder that affects millions of people worldwide, causing cognitive decline, memory loss, and behavioral changes. While there is no cure for dementia, pharmacological treatments aim to manage symptoms and slow down disease progression. This article provides an overview of the current pharmacological options, including cholinesterase inhibitors and NMDA receptor antagonists, commonly used in dementia treatment. Additionally, it explores emerging therapies, such as anti-amyloid therapies, tau protein stabilizers, anti-inflammatory drugs, and neuroprotective agents, which hold promise for modifying the underlying pathology of dementia. Continued research in pharmacological interventions is crucial for developing more effective treatments and improving the lives of individuals affected by dementia.

Keywords: Creutzfeldt-Jakob disease; Prions; neurodegeneration; Protein misfolding; Prion propagation; Prion strains; Amyloid aggregates; Synaptic dysfunction; Neuronal death; Diagnostic strategies; therapeutic interventions

Introduction

Creutzfeldt - Jakob disease stands as a mysterious and devastating neurodegenerative disorder that has puzzled scientists and clinicians alike for decades. With its enigmatic origins and rapid progression, this disease sheds light on the intricate relationship between prions and neurodegeneration. In this article, we delve into the complexities of Creutzfeldt - Jakob disease, exploring the role of prions in its development and the broader implications for our understanding of neurodegenerative disorders [1].

At the heart of CJD lies an enigmatic agent known as a prion. Unlike typical infectious agents such as viruses or bacteria, prions lack nucleic acids and consist solely of misfolded proteins. The conversion of normal cellular prion protein into its misfolded and pathogenic isoform is a hallmark of prion diseases. This conversion leads to the accumulation of PrP^Sc aggregates in the brain, resulting in the characteristic spongiform appearance seen under a microscope.

The connection between prion misfolding and neurodegeneration has not only expanded our understanding of protein misfolding diseases but has also challenged conventional ideas of infectious agents. This disease paradigm has far-reaching implications not only for CJD but also for other neurodegenerative disorders such as Alzheimer's and Parkinson's diseases, where protein misfolding and aggregation play a central role [2].

Understanding creutzfeldt - jakob disease

Creutzfeldt - Jakob disease is a rare and fatal disorder affecting the brain and nervous system. It belongs to a group of diseases called transmissible spongiform encephalopathies, characterized by the accumulation of abnormal proteins in the brain. The disease manifests with a range of neurological symptoms, including memory loss, personality changes, muscle stiffness, twitching, and eventually severe mental impairment, leading to a vegetative state and death. The disease progresses rapidly, often within a year of onset.

There are different forms of CJD, including sporadic, familial, and acquired variants. Sporadic CJD occurs spontaneously without any apparent cause, while familial CJD is associated with genetic mutations. Acquired CJD can result from exposure to contaminated tissues, such as eating meat from animals with bovine spongiform encephalopathy, commonly known as mad cow disease [3].

The culprit behind the disease

At the heart of Creutzfeldt - Jakob disease lies an unusual and perplexing agent: prions. Prions, short for proteinaceous infectious particles, are abnormal forms of a normal cellular protein called prion protein. What makes prions unique is their ability to induce normal PrP proteins to adopt the misfolded, infectious conformation, leading to a chain reaction of protein misfolding and aggregation in the brain. In the context of CJD, these misfolded prion proteins accumulate in brain tissue, forming plaques that disrupt normal brain function. The exact mechanism by which prions cause this misfolding and aggregation remains an active area of research. Their role in other neurodegenerative disorders, such as Alzheimer's and Parkinson's diseases, has also garnered attention, suggesting potential commonalities in protein misfolding mechanisms across diseases [4].

The prion hypothesis and beyond

The discovery of prions and their association with Creutzfeldt - Jakob disease led to the formulation of the prion hypothesis, which challenged conventional wisdom about infectious agents. Unlike viruses or bacteria, prions lack genetic material and rely solely on protein misfolding for propagation. This groundbreaking concept has transformed our understanding of disease transmission and protein dynamics. The prion hypothesis also opened doors for investigating other neurodegenerative disorders with a focus on protein misfolding. Researchers are exploring whether similar mechanisms underlie diseases like Alzheimer's, Parkinson's, and amyotrophic lateral sclerosis, as well as how these insights might pave the way for innovative therapeutic strategies [5].

***Corresponding author:** Kaokaf Gonda, Department of Psychiatry and Biobehavioral Science, UCLA School of Medicine, USA, E-mail: kaokaf.gonda@gmail.com

Received: 02-Sep-2023, Manuscript No: dementia-23-111565, **Editor Assigned:** 04-Sep-2023, pre QC No: dementia-23-111565 (PQ), **Reviewed:** 18-Sep-2023, QC No: dementia-23-111565, **Revised:** 21-Sep-2023, Manuscript No: dementia-23-111565 (R), **Published:** 28-Sep-2023, DOI: 10.4172/dementia.1000173

Citation: Gonda K (2023) Creutzfeldt - Jakob Disease from Prions to Neurodegeneration. J Dement 7: 173.

Implications for treatment and future directions

Although there is no cure for Creutzfeldt - Jakob disease, understanding the role of prions in its development has raised hopes for potential treatments. Researchers are exploring various strategies to inhibit prion replication, block protein misfolding, or promote the clearance of abnormal proteins from the brain. These approaches could potentially slow down or halt disease progression in the future. Furthermore, the lessons learned from studying prions in CJD have broader implications for the field of neurodegeneration. The interconnectedness of protein misfolding and aggregation across different diseases suggests that targeting these common mechanisms could lead to innovative therapies that transcend individual disorders.

Discussion

Creutzfeldt-Jakob disease is a rare, degenerative, and ultimately fatal brain disorder that falls under the broader category of prion diseases. Prion diseases are caused by misfolded proteins known as prions, which have the unique ability to induce other normally folded proteins to adopt the abnormal prion conformation. This process leads to the accumulation of misfolded proteins, primarily in the brain, and results in neuronal damage and neurodegeneration [6].

Prions and protein misfolding

Prions are primarily composed of a misfolded isoform of a cellular protein called the prion protein. PrP exists in two conformations: a normal, harmless form and a misfolded, infectious form. The misfolded form has a different three-dimensional structure that is rich in beta-sheet secondary structures, making it more resistant to degradation by cellular mechanisms.

The exact mechanism by which prions induce the misfolding of normal PrP^C into the abnormal PrP^Sc is not completely understood, but it is believed to involve a template-driven process. The presence of PrP^Sc acts as a seed that catalyzes the conversion of PrP^C into its misfolded counterpart. This conversion process leads to the accumulation of PrP^Sc, forming aggregates and amyloid plaques in the brain [7].

Creutzfeldt - jakob disease and neurodegeneration

In Creutzfeldt-Jakob disease, the accumulation of misfolded PrP^Sc in the brain leads to a cascade of events that ultimately result in neurodegeneration. As these prion aggregates build up, they disrupt normal cellular processes, including protein degradation pathways and cellular signaling. This disruption contributes to the dysfunction and death of neurons, leading to the characteristic clinical features of the disease, such as cognitive decline, behavioral changes, and motor deficits.

The neurodegenerative process in CJD involves several mechanisms

Neuronal damage: Misfolded prions directly damage neurons, leading to their dysfunction and eventual death. This contributes to the progressive neurological symptoms observed in CJD patients [8].

Inflammation: The presence of misfolded proteins triggers an inflammatory response in the brain, involving microglia and astrocytes. While inflammation is a normal defense mechanism, chronic and excessive inflammation can exacerbate neurodegeneration.

Synaptic dysfunction: Prion aggregates disrupt synaptic function, impairing communication between neurons. Synaptic dysfunction is a hallmark of neurodegenerative diseases and contributes to cognitive and motor deficits.

Cellular stress: Cells under stress activate cellular stress response pathways. In the case of CJD, the presence of misfolded prions triggers stress responses, which can further contribute to cell dysfunction and death.

Spread of prions: Misfolded prions can propagate throughout the brain by inducing the conversion of normal PrP^C into PrP^Sc. This leads to the progressive spread of pathology and the involvement of multiple brain regions [9].

Diagnosis and treatment:

Diagnosing CJD involves a combination of clinical assessment, imaging techniques, cerebrospinal fluid analysis, and, in some cases, brain biopsy. Unfortunately, there is no cure for CJD, and treatment focuses on managing symptoms and providing supportive care [10].

Conclusion

Creutzfeldt - Jakob disease serves as a poignant reminder of the complex and intricate relationship between prions and neurodegeneration. It devastating impact on individuals and families underscores the urgency of unraveling the mysteries of prion diseases and developing effective treatments. As our understanding of prions expands, so does the potential for groundbreaking discoveries that could revolutionize our approach to treating neurodegenerative disorders, offering hope for a better future for those affected by these devastating conditions.

Conflict of Interest

None

Acknowledgment

None

References

1. Adler G, Lawrence BM, Ounpraseuth ST, Asghar-Ali AA (2015) A Survey on Dementia Training Needs Among Staff at Community-Based Outpatient Clinics. Educational Gerontology 41: 903–915.
2. Bokshan SL, Han AL, DePasse JM, Eltorai AEM, Marcaccio SE, et al.(2016) Effect of Sarcopenia on Postoperative Morbidity and Mortality After Thoracolumbar Spine Surgery. Orthopedics 39:e1159–64.
3. Abdelaziz M, Samer Kamel S, Karam O, Abdelrahman (2011) Evaluation of E-learning program versus traditional lecture instruction for undergraduate nursing students in a faculty of nursing. Teaching and Learning in Nursing 6: 50-58.
4. Warrick N, Prorok JC, Seitz D (2018) Care of community-dwelling older adults with dementia and their caregivers. CMAJ 190: E794–E799.
5. Skovrlj B, Gilligan J, Cutler HS, Qureshi SA (2015) Minimally invasive procedures on the lumbar spine. World J Clin Cases 3:1–9.
6. Allen M, Ferrier S, Sargeant J, Loney E, Bethune G, et al. (2005) Alzheimer's disease and other dementias: An organizational approach to identifying and addressing practices and learning needs of family physicians. Educational Gerontology 31: 521–539.
7. Surr CA, Gates C, Irving D, Oyebode J, Smith SJ, et al. (2017) Effective Dementia Education and Training for the Health and Social Care Workforce: A Systematic Review of the Literature. Rev Educ Res 87: 966-1002.
8. Ruiz JG, Mintzer MJ, Leipzig RM (2006) The Impact of E-Learning in Medical Education. Acad Med 81(3):207-212.
9. Reeves S, Fletcher S, Loughlin C, Yim A, Patel KD (2017) Interprofessional online learning for primary healthcare: findings from a scoping review. BMJ Open 7: 168-172.
10. Kafil TS, Nguyen TM, MacDonald JK, Chande N (2018) Cannabis for the treatment of ulcerative colitis. Cochrane Database Syst Rev 11:129-154.