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Causes and Imaging of Creutzfeldt–Jakob Disease

Yamaguche Kawa*

Department of Neurology, University of Calgary, Alberta, Canada

Commentary

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***For Correspondence:** Yamaguche Kawa, Department of Neurology, University of Calgary, Alberta, Canada.

E-mail: kawa.yamaguche@a7g.edu

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DESCRIPTION

Creutzfeldt–Jakob Disease (CJD) also known as subacute spongiform encephalopathy or neurocognitive dysfunction caused by prion disease, is a deadly degenerative brain condition. Early signs may include visual abnormalities, behavioural changes, memory issues, and poor coordination.Dementia, uncontrollable movements, blindness, weakness, and coma are some of the later signs. 70% of patients pass within one year after receiving a diagnosis. After the German neurologists Hans Gerhard Creutzfeldt and Alfons Maria Jakob, Walther Spielmeyer first coined the term "Creutzfeldt-Jakob disease" in 1922. A prion is a class of aberrant protein that causes CJD. Misfolded proteins called infectious prions can alter the way that regularly folded proteins fold. Approximately 85% of CJD cases are caused by unknown factors, while 7.5% are caused by autosomal dominant inheritance. Spread may also occur when exposed to brain or spinal tissue from an infected person. There is no proof that sporadic Creutzfeldt-Jakob disease can spread through regular contact or blood transfusions, notwithstanding the possibility in variant Creutzfeldt-Jakob disease. Diagnosis entails eliminating any more probable reasons. The diagnosis may be supported by an electroencephalogram, a spinal tap, or magnetic resonance imaging.

Since the majority CJD is not specifically treated. Clonazepam or sodium valproate may be used to treat uncontrollable movements, while opioids may be used to treat pain. One in a million persons are affected by CJD each year. Onset normally occurs at the age of 60. The illness was initially explained in 1920.This form of transmissible spongiform encephalopathy is categorized as such. One in ten cases of prion disease is inherited CJD. Sporadic Creutzfeldt-Jakob Disease (CJD) is distinct from Variant Creutzfeldt-Jakob Disease (vCJD) and bovine spongiform encephalopathy (mad cow disease).CJD is a form of TSE (Transmissible Spongiform Encephalopathy), which is brought on by prions. Proteins that have been improperly folded are known as prions, and they can develop in Central Nervous System (CNS) neurons. They are believed to interfere with signalling pathways, harming neurons and causing degeneration that gives the diseased brain its spongiform appearance.

The CJD prion is harmful because it encourages natural prion protein to refold into the diseased state. Protein molecules that are misfolded will multiply exponentially, which results in a significant amount of insoluble protein in the affected cells. This accumulation of improperly folded proteins impairs neuronal cell function and results in cell death. Misfolding of the predominately alpha helical regions into beta pleated sheets can result from mutations in the prion protein gene. Because of this alteration in structure, the protein can no longer be digested. Upon prion transmission, the damaged proteins enter the brain and cause additional prion protein molecules to misfold in a self-sustaining feedback loop. These neurodegenerative conditions are frequently referred to as prion diseases.

Additionally, individuals who have a mutation in the PRNP gene, which codes for the prion protein, are susceptible to CJD. Approximately 5–10% of CJD cases involve this. The misfolding of the prion protein is a process that is thought to happen in rare cases as a result of aging's impact on cellular machinery, which explains why the disease frequently manifests later in life. According to an EU research, "87% of cases were sporadic, 8% of them genetic, 5% of them iatrogenic, and less than 1% of them variant.During a medical examination, brain imaging may be done to both rule out other explanations and to gather data to support a diagnosis. Both the appearance and the sensitivity and specificity of imaging findings might vary. While imaging is less important in the diagnosis of CJD, some brain MRI findings may occasionally appear before the beginning of clinical symptoms. The most effective imaging technique for CJD-related alterations is brain MRI. Diffuse-weighted imaging sequences are the most sensitive MRI sequences.