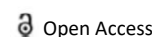




Commentary



Transmission of Creutzfeldt–Jakob Disease and its Symptoms

Jaly Cela*

Department of Paediatric Cardiology, University Hospital Münster, Münster, Germany

Description

Creutzfeldt-Jakob disease (CJD), also known as sub-acute spongiform encephalopathy or prion neurocognitive disorder, is an invariably fatal degenerative brain disorder. Early symptoms include memory problems, behavioral changes, poor coordination and vision disturbances. Later symptoms include dementia, involuntary movements, blindness, weakness, and coma. About 70% of people die within a year of diagnosis. The name Creutzfeldt–Jakob disease was introduced by Walther Spielmeyer in 1922 after the German neurologists Hans Gerhard Creutzfeldt and Alfons Maria Jakob.

Creutzfeldt-Jakob disease is caused by a type of abnormal protein known as a prion. Infectious prions are misfolded proteins that can cause normally folded proteins to also misfold. About 85% of Creutzfeldt-Jakob disease cases occur for unknown reasons, while about 7.5% of cases are inherited in an autosomal dominant manner.

Transmission

The defective protein can be transmitted by contaminated human brain products, corneal grafts, dural grafts or electrode implants, and human growth hormone.

May be familial Creutzfeldt-Jakob disease; or it may occur without clear risk factors. In the familial form, a mutation in the gene for PrP, PRNP, occurred in this family. All types of Creutzfeldt-Jakob disease are transmissible regardless of how they occur in a person.

It is thought that humans can contract a variant form of the disease by eating food from animals infected with bovine spongiform encephalopathy (BSE), a bovine form of Transmissible spongiform encephalopathies, also known as mad cow disease. However, in some cases it can also cause sporadic Creutzfeldt-Jakob disease.

Cannibalism has also been implicated as a transmission mechanism for abnormal prions, causing the disease known as kuru, which once occurred primarily among

ARTICLE HISTORY

Received: 15-Nov-2022, Manuscript No. JCMEDU-22- 82538;
Editor assigned: 18-Nov-2022, Pre-QC No. JCMEDU-22- 82538 (PQ);
Reviewed: 02-Dec-2022, QC No. JCMEDU-22- 82538;
Revised: 09-Dec-2022, Manuscript No. JCMEDU-22- 82538 (R);
Published: 16-Dec-2022

the women and children of the Forä people of Papua New Guinea, who formerly practiced funerary cannibalism. While the men of the tribe ate the muscle tissue of the deceased, the women and children consumed other parts such as the brain and were more likely than the men to contract kuru from the infected tissue.

Prions, the infectious agent of Creutzfeldt-Jakob disease, cannot be inactivated by standard surgical sterilization procedures. The World Health Organization and the US Centers for Disease Control and Prevention recommend that used instrumentation in such cases be destroyed immediately after use; without destruction, it is recommended to use thermal and chemical decontamination in combination for processing instruments that come into contact with highly infectious tissues. Thermal depolymerization also destroys prions in infected organic and inorganic matter, as the process chemically attacks the protein at the molecular level, although more effective and practical methods involve destruction with combinations of detergents and enzymes similar to biological washing powders.

Symptoms

The first symptom of Creutzfeldt-Jakob disease is usually include rapidly progressive dementia leading to memory loss, personality changes and hallucinations. Myoclonus (jerking movements) typically occurs in 90% of cases, but may not be absent at the onset. Other common features include anxiety, depression, paranoia, obsessive-compulsive symptoms, and psychosis. This is accompanied by physical problems such as slurred speech, dysfunction of balance and coordination (ataxia), changes in gait and stiff posture. In most people with Creutzfeldt-Jakob disease, these symptoms are accompanied by involuntary movements. The duration of the disease varies greatly, but sporadic (non-hereditary) Creutzfeldt-Jakob disease can be fatal within months or even weeks. Most affected people die six months after the first symptoms appear; often from pneumonia due

to impaired cough reflexes. About 15% of people with CJN survive for two years or more.

The symptoms of Creutzfeldt-Jakob disease are caused by the progressive death of brain nerve cells, which is associated with the accumulation of abnormal prion proteins

that form in the brain. When brain tissue from a person with Creutzfeldt-Jakob disease is examined under a microscope, many small holes can be seen where nerve cells have died. Parts of the brain may resemble a fungus where prions have infected areas of the brain.