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Mini Review

Cruetzfeldt-Jakob Disease and Mode of Transmission

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Abstract

Cruetzfeldt-Jakob disease (CJD) exists in inherited, acquired (variant and iatrogenic), and spontaneous (sporadic) forms. Although iatrogenic and variant forms of CJD usually affect relatively young people, all forms may affect elderly persons, especially sporadic CJD. Sporadic CJD is a rare cause of dementia among middle aged and elderly persons and typical cases are clinically fairly distinct from more common forms. However, clinical diagnosis can be a challenge for those who are not experienced with the disease. Certain investigations can be very helpful although many cases of CJD (especially sporadic CJD) are not thought to be acquired illnesses, there is still a potential for onward transmission and certain precautions are necessary to protect public health. Creutzfeldt-Jakob disease is an illness that has had major medical, media and political impact, despite its rarity, essentially because of its potential transmissibility with one dorm of CJD being a zoonosis. Much attention has been focused on variant CJD but the most common form is sporadic CJD. This review article focuses on the general information about CJD, especially its mode of transmission and prevention because just as they say "prevention is better than cure", it is advisable to be informed and keep at watch to avoid getting them if not through inheritance or sporadically.

Creutzfeldt-Jakob Disease and its Mode of Transmission

Cruetzfeldt-Jakob Disease (CJD) and its variants belong to a broad group of human and animal disease known as transmissible spongiform encephalopathies (TSEs). The name was derived from the spongy holes, visible under microscope, that develop in affected brain tissues. It is a rare degenerative, invariably fatal brain disorder that occurs in elderly people of 50 years and above.

Causative Agent

The cause of Cruetzfeldt-Jakob disease appears to be abnormal versions of a kind of protein called Prion. Normally, these proteins are harmless but when they're misshapen, they become infectious and can harm normal biological processes.

Mode of Transmission

The risk is low. The disease is not contagious, there are three major ways it can be transmitted or it can develop;

1. Sporadically: Most people with classic CJD develop the disease for no apparent reason.
2. By Inheritance: In the United States, about 5-10% of people with CJD have a family history of the disease or test positive for a genetic mutation associated with CJD. This type is referred to as familial CJD.
3. By contamination: A small number of people develop CJD after being exposed to infected human tissue during a medical procedure, such as a cornea or skin transplant. Also, because standard sterilization methods do not destroy abnormal prions, a few people have developed CJD after undergoing brain surgery with contaminated instruments. Some with variant CJD, they get CJD primarily by eating beef infected with mad cow disease (Bovine spongiform encephalopathy or BSE).

Symptoms

Initial signs and symptoms include;

1. Personality changes
2. Depression
3. Anxiety
4. Difficulty in speaking and swallowing
5. Sudden jerky movement
6. Insomnia
7. Blurred vision
8. Impaired thinking

As the disease progresses, mental symptoms worsen. Most people eventually lapse into a coma. Heart failure, respiratory failure, pneumonia or other infections are generally the cause of death. Death usually occurs within a year in people with the rarer variant CJD, psychiatric symptoms may be more prominent in the beginning with dementia (loss of the ability to think, reason and remember) developing later in the illness. In addition, this variant CJD affects people at a younger age than the classic CJD does and it appears to have a slightly longer duration of 12 to 14 months.

Diagnosis

Currently there is no single diagnostic test for CJD. A neurological examination will be performed and the doctor may seek consultation with other physicians. Standard diagnostic test will include a spinal tap to rule out more common causes of dementia and an electroencephalogram (EEG) to record the brain's electrical pattern which is used to show the abnormality in CJD. The only way to confirm diagnosis of CJD is by brain biopsy or autopsy.

Treatment

Researchers have tested many drugs, including amantadine, steroids, acyclovir, antiviral agents and antibiotics but none worked. Opiate drugs can help relieve pain, clonazepam and sodium valproate helps to relieve myoclonus. At later stages of the disease, changing the person's position can help to keep him or her comfortable and helps to prevent bedsores. A catheter can be used to drain urine if the individual cannot control bladder function and intravenous fluids and artificial feeding also may be used.

Prevention

a. Never donate blood, tissue or organs if they have suspected or confirm CJD.
b. Sterilization.
c. Cover cuts and abrasions with water proof dressing if the patient is CJD positive.
d. Wear surgical gloves when handling patient's tissue and fluids.
e. Avoid cutting or sticking themselves with contaminated blood or tissue.
f. Use face protection.
g. Ensure that proper personal hygiene is observed.

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