Creutzfeldt jakob disease: causes, symptoms and treatment

One of the diseases that has severely impacted the lives of people these days is Creutzfeldt-Jakob disease (CJD) which is a condition that is fatal and rare which has impact on the brain of human being. CJD causes damage to the brain that gets worsened with the passage of time. The patients diagnosed with the disease die within one year as people become immobile (Solassol et al., 2006). CJD is usually caused by an abnormal protein known as prion. Before prion is understood, proteins' function for body must be clarified. For every human being, proteins are important for life and are present in body of all living things. Every part of human's body has good composition of protein such as muscles, hair, fingernails, bones, skin, blood, body organs and eyes. After water, protein is the second most important constituent of body (Belay et al., 2003).

According to LiveScience Staff (2012), protein is mainly used for building, maintenance and repairing of body tissues; it is present in various forms which perform many jobs in human body and the structure of each protein determine its function. The building blocks of proteins are long strings of amino acids which get folded and curled into complex three dimensional shapes which allow proteins to perform their job. Dr. Stanley B. Prusiner from University of California i. e. San Francisco purified an agent which was infectious made of unique kind of protein and referred to it as " prion" in 1982; Nobel Prize in Physiology or Medicine was awarded to him in 1997 for discovery of Creutzfeldt-Jakob Disease (CJD) that was mainly caused by prions.

Prions get accumulated in brain at much higher levels that cause permanent damage to the nerve cells which cause various neurological symptoms.

These infectious particles are different in behavior from conventional bacteria and viruses which cannot be destroyed by exposing to heat and radiation. Even antiviral and antibacterial medicines have no effects on prions; hence, presently, there is lack of valuable treatment for CJD (Belay et al., 2003).

Understanding Creutzfeldt-Jakob Disease (CJD)

Basically, CJD is a very rare form of deadly form of dementia which can spread fastly in the body from brain. Prions are mostly found in brain and are harmless; when they are not given proper shapes, they may have effects that can be devastating which can even attack brain, kill its cells and create holes or gaps in tissue of brain (Ironside, 2009). Prion diseases have been found in both animals and humans; such diseases were in news of mid 1980s along with Bovine Spongiform Encephalopathy (BSE) epidemic which was referred to as Mad Cow disease, it is a cattle disease. In humans, it is known as CJD. The disease is found in about one to two people among million people worldwide.

Types of CJD

There are basically two types of CJD i. e. Classic CJD and Variant CJD (vCJD). The three types of classic CJD are Sporadic CJD, Familial or genetic CJD and Iatrogenic or Acquired CJD.

Sporadic CJD

More than 90% of people diagnosed with CJD are suffering from this type of CJD and it is mainly found in people aged in between 45 to 75. There are no specific symptoms for this CJD (Shah et al., 2009); however, some early

symptoms such as depression may be present but it can quickly progress into confusion and problems of memory which is commonly observed in dementia, followed by loss of balance and coordination along with progressive blindness (Piouti et al., 2012).

Familial or genetic CJD

Familial or genetic CJD is one of the rare genetic condition in which the person's genes are inherited from one parent who causes the protein to get mutated into a prion in later stages of life that trigger the symptoms of CJD. In other words, it appears in families that have abnormal gene. About 7% of cases have this type of CJD. In order to diagnose it, blood test is done; person having this abnormal gene has 50% chance of transmitting it to children. The other rare form of this CJD are Fatal Familial Insomnia (FFI) and Gerstmann-Sträussler-Scheinker (GSS); both of these are rarely found in people (Brand et al., 2006). It occurs in people who have age of 50 years.

Iatrogenic or Acquired CJD

This type of CJD is rarely found in people as it is found in those people who get CJD while accidental transmission that takes place during a medical procedure like corneal grafts, instruments in neurosurgery, human pituitary hormone therapy or human dura-mater grafts (Mayo Clinic Staff, 2010).

Variant CJD

People having average age of 28 years are found to be suffering from Variant CJD. It is usually developed from eating of beef that was infected with BSE. It can even occur with blood transfusion in which variant CJD is transmitted

from one person to other. Once the symptoms are identified, the disease spreads quickly in the body (Tattum et al., 2010).

Signs and Symptoms of CJD

In many of the cases, classic CJD looks similar to various other dementias; the disease comes on fastly and then declines the ability of thinking as symptoms start appearing. Some of the most commonly found symptoms are that the person may have swings in mood, problems related to memory, interest absence and not behaving like oneself, having difficulty while walking such as keeping balance, dementia rapid progressions such as loss of memory and other thinking capabilities, problems in vision such as blindness, stiffening of limbs, twitching or jerks in muscles, feeling of clumsiness, speaking problem such as slurred speech, difficulty in swallowing and akinetic mutism in which person can move eyes and appear to be alert but can neither move voluntarily or speak (Espinosa, Bensalem-Owen, & Fee, 2010; Sikorska et al., 2004).

However, people who are in CJD's later stages face problems such as losing awareness as it can be seen in various neurological examinations. While the disease in the early stages, CJD patients are scared and it can be a distressing situation for them; it is usually associated with hallucinations that are visual which means seeing things that are not present there which might develop uncomfortable feeling in them (Andrews, 2012).

People who are suffering from sporadic CJD usually live than twelve months after the appearance of signs and symptoms; the maximum life is two years. Before death, some people fall into condition of coma; the main reasons of

death are heart failure, pneumonia or respiratory failure such as breathing problem (Rossetti & Dunand, 2007). Among the most common symptoms found in Variant CJD people are changes in behavior, depression, withdrawal from social gatherings, difficulty in walking, dementia that is progressive, unable to do movement or speak and pain and odd kind of sensation in limbs or face (LiveScience Staff, 2012).

Causes of CJD

According to studies of Ironside (2009) and Tattum et al. (2010), CJD is caused by slow virus or other small organisms. The agent that causes this disease has several characteristics which are different from viruses and bacteria's. The organisms are difficult to kill as they don't have genetic information on them in the form Nucleic acids such as DNA or RNA and has long period of incubation before symptoms are made visible. However, it has been found that it is mostly caused by prion proteins that can occur in both normal and infectious form. The normal forms have same shapes as that of amino acids but infectious ones differ in their shapes from normal proteins. Once they appear, abnormal ones start to get aggregated which start affecting the brain. Only 5-10% cases are inherited ones that arise either from mutation or changes in gene which are responsible for controlling the formation of normal prion proteins.

Diagnosis of CJD

CJD is difficult to diagnose especially when it is in its initial stages; there is no specific test available for diagnosing in living person. Only way to make sure that person had CJD is by examining the tissue of brain after his death. However, there are some other tests and procedures can be used for

diagnosis of the disease such as medical history examination as it will help doctors in learning the person's symptoms and signs when they get started as CJD gets spread quickly; Magnetic Resonance Imaging (MRI) in which brain's picture is taken to identify the difference between various types of CJD; Computerized Tomography (CT) scan in which brain's picture will be taken to diagnose the disease; Puncture of Lumbar which means taking fluid form the spine of a person by using syringe and a needle, it is done to identify the infections of brain; blood tests to identify if there is any chance of genetic CJD; Electroencephalogram (ECG) in which electrical activity of brain is measured and brain autopsy in which tissue of brain is examined after person's death (Puoti et al., 2012; Shah et al., 2009).

Treatment for CJD

Up till now, there has been no success in developing right treatment for CJD. The researchers have done numerous tests with many drugs such as steroids, antiviral agents, antibiotics, amantadine, acyclovir and many more but they have not been able to find perfect cure for this disease (Puoti et al., 2012). Some studies are still in progression to develop appropriate treatment for patients but none of them have been successful in benefiting the human beings.

The treatment for CJD that is being followed by various doctors is aimed at alleviation of symptoms and trying to make the individuals as comfortable as the experts can do so that they have the will power of fighting against the disease. Some of the opiate drugs can relieve pain when people suffer from it but the drugs such as Sodium Valproate and Clonzaepam definitely help in relieving of myoclonus. When the disease is in later stages, position of

person is changed frequently so that he gets comfortable and bedsores can be prevented. For draining urine, a catheter can be used as it helps in controlling the function of bladder and artificial feeding can also be used.

Precautions for CJD

Although there are chances that CJD gets transmitted by being careless during medical procedures but still people need to take care of various things that can help them in remain protected from this disease. CJD is not a contagious disease that can be transmitted either by social or sexual contact or via air or feeding, touching or even taking care of person suffering from CJD at home. some of the basic precautionary measures that need to be followed are washing hands either before eating or drinking, covering wounds or cuts with bandages that are waterproof, protecting face and hands from being exposes to blood or fluids of body of person who is affected by this disease and taking special care in blood transfusions so that CJD infected person does not give his blood.

In order to avoid variant CJD, beef from selected countries should be eaten; countries that have high risk of TSE are more vulnerable to risk. The only options available for avoiding this type of CJD are either to eat beef from countries that emphasize on strict regulations on its quality and avoid eating parts of cattle that have high risks such as spinal cord, intestines, brain and eyes.

Conclusion

Creutzfeldt – Jakob disease (CJD) is among the fastest growing disease that is being faced by many people worldwide. It has become important for

researchers to identify right course of treatment for this disease so that patients can get full recovery. In order to make sure that people who have genetic CJD are given appropriate treatment before symptoms start to become visible, it is important that people get their blood tests done at regular intervals. When any of the signs and symptoms is observed in people, it is advised that they contact a doctor immediately who can ensure that right tests are done for identification of this disease.

Still, the researchers have to do extensive work for identifying the main causes of CJD along with developing a remedial solution. Among all the available options, the best one is to contact any well-known doctors who are specialist in neurology who will take the patient in right treatment method. Hence, everyone needs to make sure that they have complete information about this disease so that they can face it with determination and strong will power.