Correspondence:

Bovine spongiform encephalopathy (mad cow disease)

During 1985-1986, farmers in the United Kingdom (UK) noticed that their cattle are suffering from an unknown disease characterized by change in behaviour, inability to walk, stand on the ground and finally death. Autopsy of these animals showed spongiform degeneration of the brain tissue and so the scientists named this disease as bovine spongiform encephalopathy (BSE) or mad cow disease (MCD).

After years of research on this disease, Stanley Prusiner found that the disease is spreading even after destroying all the genetic material i.e., deoxy ribonucleic acid (DNA) and ribonucleic acid (RNA) from the serum of the infected animals and its failure to spread after selectively destroying proteins. This experiment proved that the disease is spread by a protein particle and he named this new and interesting infectious protein as “prion”. Stanley Prusiner was awarded with Nobel Prize in 1997 for the discovery of this new type of infecting agent.

Paralleling this epizootic mad cow disease in cattle, there was a rise in number of people suffering from Creutzfeldt-Jakob disease (CJD) like disease in UK. It differs from the classical CJD by its occurrence in young people (mean age at death = 29 years) compared to the older age (65-79) years for classical CJD, prolonged course of the disease, higher mortality and higher incidence of myoclonus, depression and anxiety. This new disease was named as new variant Creutzfeldt-Jakob disease (vCJD).

Cattle-to-cattle transmission occurs mainly by use of infected cattle bones and meat in the preparation of cattle feed. There are no proven cases of MCD spread by direct cattle-to-cattle contact.

Prions the causative agents of MCD are found abundantly in nervous tissue of infected cattle. Cattle-to-human transmission of vCJD occurs mainly by eating of infected beef. Other ways of spread of infection include use of vaccines grown on infected cow’s tissue, and gelatins prepared from cattle. In December 2003, death of an adult from vCJD who it is believed acquired the infection from blood transfusion has also been reported.

Corneal transplantation, dura mater transplantation and use of human growth hormone are the other important methods of transmission of the disease. Corneal transplantation is of special concern because of its wide spread usage. Other theoretically possibly methods of infection include: through EEG electrodes, seminal fluid, organ donation, in-utero transmission from mother-to-child and, via breast milk.

MCD is a gradually progressive disease that eventually kills the animal. The usual life-span after appearance of the symptoms varies from 6 months to 2 years. The disease starts manifesting only in cattle which are above 30 months of age. Gradual change is observed in the behaviour of the cattle; loss of coordination and difficulty of performing simple movements like standing and walking (downing cow),
decreased weight in spite of normal appetite, decreased milk production and death ensue.

The symptoms of vCJD in humans are similar to that of cattle. Incubation period is 1.5-2 years and patients die 6-12 months after appearance of symptoms. In the affected individuals, there is a definite change in personality, emotional disturbance, difficulty in movement, myoclonus, coma and death.

MCD is suspected in cattle by the clinical sign and symptoms. Definitive diagnosis is obtained only after histopathological examination of autopsied brain tissue. Live screening tests are under research and not approved for screening. There are no diagnostic tests which can detect the vCJD before the onset of symptoms. Magnetic resonance imaging (MRI) and electroencephalogram (EEG) show some non-specific changes in these patients. Definitive diagnosis is obtained only after histopathological examination of autopsied brain tissue.\(^8\)

There is no specific treatment for MCD or vCJD. Symptomatic medication is prescribed to reduce the symptoms and comfort the patient. Currently available treatment does not change or delay the death.

MCD was first observed in United Kingdom in 1985, and was specifically diagnosed in 1986. By 1990, there were some 14,000 confirmed cases. The epizootic reached its peak in 1992-93 with a incidence 1000 cases per week. Within a few years the disease spread to various countries. The following table gives the number of identified cases of BSE in various countries and the vCJD diseases.

**Implications to India**

India exports about 50,000 metric tonnes of bovine meat to over 60 countries annually, earning valuable foreign exchange of nearly ₹ 50 billions. There are no reported cases of MCD/BSE or CJD from India till date. World Organization for Animal Health (WOAH), provided “the most secure status for the deadly MCD” to India in its 78th General conference 2010.\(^8\)

Loss of cattle by MCD or as a part of the eradication program is associated with financial burden to many farmers which may lead to the migration of farmers from villages to cities which could impair the social structure of the society and it will be extra burden to the urban government to deals with increased migrants.

The dependent children are orphaned by loss of a caring person in a family with vCJD, which could have serious consequences in future society. The vCJD disease mainly affects younger generation leading to loss of nations work force leading to decreased Gross national product (GNP) of the nation.

We have learned from the history that prions can cross the species barrier and cause disease in different species which may lead to loss of biodiversity. As beef is one of the largest consumed food in the world and there is wide spread export and import between different countries, vCJD had the potential to become a pandemic, if proper international policies were not implemented in this export and import business. Before establishment of disease in 1986 it was estimated that more than 80,000 cattle already entered the human food chain.

Even though MCD is not a public health problem as of now in India, as the infectious agent can spread from cattle to man by crossing the species barriers, similar types of inter-species spread can occur and MCD has the potential to emerge as a public health problem in India. As there is no specific treatment or vaccination available for MCD, prevention appears to be the only way to control this disease.

Prevention of spread of infection from cattle to man is the most important step in the control
of vCJD in humans. This is achieved by culling of suspected animals, and banning of high risk material. Surveillance by regular inspections of slaughtered animal samples, banning of used cattle products in vaccine preparation and gelatin preparation and for transplantation (bovine valve graft) are other important measures to decrease cattle to human spread.

Prevention of spread from humans to humans can be achieved by discarding cornea, blood or other organs from vCJD patient, prohibiting seminal fluid donation by persons who are suspected to be exposed to vCJD, among others. Iatrogenic transmission by improperly disinfected instruments is a theoretical possibility which can be prevented by proper disinfection at high temperatures and with strong disinfecting agents.

Political commitment is also very important in preventing the disease from spread within the nation and intercontinental spread.

REFERENCES


S. Kamalakar,¹
T. Javanthi.²

¹Research coordinator-Veterans Affairs Medical Center, Kansas City, MO, USA.
²Chalmeda Anand Rao Institute of Medical Sciences, India.