Mad Cow Disease
Bovine Spongiform Encephalopathy

Ashutosh Chachra, Deepti Narang and Raman Narang

Mad Cow Disease, or bovine spongiform encephalopathy (BSE) is one of the important zoonotic diseases, which can cause fatality in humans eating infected beef. This disease is related to another serious disease of sheep called 'scrapie' and Creutzfeldt-Jacob disease in humans. In the past few years, this disease was in the news as a result of a major epidemic in British cattle herds (see Box 1) and over 1,60,000 cattle had died in UK by the end of January 1997.

Etiology of BSE

BSE and other related diseases like scrapie are caused by an unconventional proteinaceous infective agent called 'prion' (see Box 2). Prion caused diseases are also known in sheep, mink, muskdeer, and humans. The prion of scrapie, to which the prion causing BSE is closely related, is not destroyed by usual viricidal procedures. It can withstand boiling for 30 minutes, rapid freezing and rapid thawing, and even exposure to chemical agents like ether and 20% formalin. Moreover, prions are not antigenic, so no antibodies are evoked against them and thus no humoral immunity is developed against such infections.

Transmission of BSE

The story of the transmission and spread of BSE dates back to 1981–82, when cattle in UK were fed with protein from bone meal and meat meal. It is thought that these protein meals contained prions from scrapie infected sheep, because discarded remains of sheep, including brains, were used to prepare cattle feed. After a long incubation period of 22 months, which is not uncommon in prion diseases, BSE first surfaced in 1985.
Box 1. Hue and Cry in the United Kingdom

Out of 12 million cattle in UK, over 1,60,000 became victims of mad cow disease in the past few years. Due to the presence of this deadly disease in British cattle stocks, the European Community banned the import of British beef, causing a slump in the 12 billion pound beef industry in Britain, and a large scale slaughter of infected herds.

This epizootic disease was recorded in recent years only in Great Britain and Northern Ireland. Cases have also occurred in Oman and the Republic of Ireland, in cattle imported from Great Britain, while sporadic cases have occurred in native cattle in Switzerland, Ireland, Denmark, the Channel Islands and France. A similar disease has also occurred in a small number of exotic bovines in zoos in Britain. Spongiform encephalopathy has also been reported in domestic cats. It is, however, predicted that the BSE epidemic will be largely over by 2001, although this requires action by other European countries which may have imported young cattle from UK between 1985 and 1990, before it was realized that many thousands of British cattle herds were infected.

Human infection is supposed to have been caused by eating the brain and neural tissue of cattle which contain abundant numbers of prions. The disease can be transmitted experimentally to sheep and goats. Other horizontal or vertical transmission similar to that of typical infectious diseases has not so far been reported. Similarly, milk is thought to lack infectivity, but the use of milk from BSE infected animals has, nevertheless, been banned since 1988.

Clinical Signs of BSE

The clinical signs of BSE in cattle include apprehension, change in temperament, teeth grinding, excessive sensitiveness, excessive licking, head pressing and rubbing, uncoordination of movement, blindness, tremors, falling and lying down, fetlock knuckling, circling, partial paralysis, loss of weight and reduced milk yield.

There is bilateral vacuolation of gray matter neurons (spongiosis) which can only be detected by a histopathological examination.

Diagnosis and Treatment of BSE

BSE is difficult to diagnose as it is usually confused with diseases like hypomagnesemia, nervous form of ketosis, furious form of rabies, encephalic listeriosis, lead poisoning and Downer’s cow

It is thought that BSE in cattle resulted from feeding cattle with feed made from remains of dead sheep, some of which were infected with the prion disease, scrapie.
Box 2. Prions: A Unique Type of Infectious Agent

Most infective agents, ranging from the more complex protozoans to bacteria and viruses, contain nucleic acids (DNA or RNA). Even viruses that are dependent on the host cells’ biochemical machinery to reproduce, contain nucleic acids. Prions, however, seem to contain no nucleic acid, although some scientists believe there may be minute amounts of DNA associated with infective prions. Interestingly the prion protein is actually merely a modified form of a common host encoded protein that is normally produced by the cells of the host. The host protein can change its shape, by some as yet unknown mechanism, and form tangled agglomerations that impair the functioning of nerve cells, and thus of the brain. This change of shape occasionally occurs spontaneously, or as a result of contact with the infective version of the prion protein. This latter event is an interesting example of a infective protein modifying the host proteins without any involvement of nucleic acids. The 1997 Nobel prize for Physiology or Medicine was awarded to Stanley B Prusiner for the discovery and characterization of prions (see Resonance Vol 2, No. 12, pp 108–112, 1997).

Suggested Reading


syndrome. BSE can be diagnosed on the basis of the following: clinical signs, electroencephalography, magnetic resonance imaging technique, invasive procedure of brain biopsy and biological assay.

The confirmation of the disease is done by histopathological examination of brain.

There is so far no successful specific treatment for BSE. Only the sacrifice of the animal is thought to be helpful to control the infection.

Control of BSE

A group of international experts convened by WHO have recommended the following points for controlling BSE:

- All countries should ensure the slaughter and safe disposal of animals affected with any transmissible spongiform encephalopathy, so that it does not enter any food chain, human or animal.
- All countries should review their rendering procedures to ensure that they have effectively inactivated BSE agents.
- Continuous surveillance and compulsory notification of BSE according to the recommendations of the Office of International Epizootics should be made.