

Epidemiology of Ovine Scrapie in Japan

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Abstract

During the period 1984 to 1987, postmortem brain tissues were obtained in 23 samples of pathologically confirmed ovine scrapie in Japan. Although most of them were collected from Hokkaido, some brain tissues originated from Yamagata, Fukushima and Kanagawa. Occasional incidences of temporal and spatial clustering on a non-familial basis have been reported in the past, each case of which included a small number of sheep centers in Hokkaido and Fukushima. This geographical and temporal clustering, however, did not apply to the present cases with a few exceptions. Ninety percent of those incidences indicated a familial association.

A majority of the diseased sheep were from Suffolk strain, while three cases were from Corriedale strain. It is very likely that sheep-to-sheep transmission of scrapie has taken place in Obihiro, Hokkaido, Japan.

Discipline: Animal health

Additional key words: prion, spongiform encephalopathy, unconventional virus

Although ovine scrapie has been studied much more intensively than any other unconventional virus, the mechanism of its spreading under natural conditions is not identified yet. It might spread from naturally infected sheep to un-infected sheep, though such a lateral transmission has never been reported so far in sheep or goats experimentally infected. It appears to pass from ewes to lambs, even without suckling; the contact of the lamb with the infected ewe at birth appears to be effective, because the placenta itself is highly infectious¹⁰. Transplacental infection during the perinatal period rather than oral, nasal, optic, or cutaneous infections might be a possible route of transmission, though not confirmed². The first appearance of scrapie agent in young lambs naturally infected occurred at the age of 10 to 14 months in tonsils, suprapharyngeal and mesenteric lymph nodes and intestine, suggesting infections taking place in an alimentary tract. Older sheep were infected only after a long contact with diseased animals. Incidences of the disease took place in pastures previously occupied by scrapie sheep³.

Although scrapie has a widespread distribution in

Europe, Asia and America¹¹, few reports are available for Japanese cases. The present paper reviews outbreaks of ovine scrapie in northern Japan and discusses possible lateral transmission of the disease.

Since the first outbreak of scrapie in Obihiro of Hokkaido, Japan in 1984^{5,12}, postmortem brain tissues have been attained in more than 20 cases of pathologically confirmed scrapie. Clinical records on more than 10 cases have been made available. The prevalence of scrapie has varied markedly in time and place throughout northern Japan. Since attention was drawn to the syndrome of ovine scrapie, particularly its lateral transmission in Obihiro area, more frequent diagnoses have been undertaken than before by various animal health stations in Hokkaido⁷. During the period 1984 to 1986, approximately 50 cases of incidences were reported in Hokkaido. There were several cases outside Hokkaido: i.e. one scrapie-like disease incidence in Yamagata in 1983, two pathologically confirmed cases in Yamagata and Fukushima both in 1984, and scrapie-like disease cases in Iwate, Ibaraki and Chiba in 1984. In 1987, three incidences were informed in Kanagawa: the presence of scrapie was patho-

logically confirmed in one of them through brain tissue inspections (Fig. 1). In regard to the duration of ovine scrapie disease, it is a 3- to 6-month period on an average.

It was earlier reported that occasional incidences of temporal and spatial clustering had taken place on a non-familial basis in a few sheep centers in Hokkaido and Fukushima. This geographical and temporal clustering, however, did not apply to a majority of the cases as mentioned above. It was recognized that 90% of those incidences were

familial. The diseased sheep were mostly from Suffolk strain, and three cases were from Corriedale strain. It is quite probable that sheep-to-sheep transmission of scrapie took place in Obihiro, Hokkaido, since approximately 50 cases in Suffolk and three cases in Corriedale originated from the sheep in Obihiro or those transferred from there⁹⁾.

Natural scrapie in sheep is pathologically characterized by the presence of vacuolated neurons and extensive astrogliosis. In the cases as mentioned above, the presence of amyloid plaques is rare. A



Fig. 1. Distribution of scrapie-affected sheep by prefecture, 1984 to 1987

high frequency of vacuolated neurons was seen in nucleus olivaris in medulla oblongata⁷⁾.

The scrapie agent was purified by density-gradient sedimentation and, more effectively, in the presence of specific detergent¹³⁾. Scrapie infectivity could be easily sedimented by ultracentrifugation. Merz et al. demonstrated an amyloid-like two- or four-stranded fiber in the sediment, which increases in quantity with infectious titer using mice⁴⁾. Such scrapie-associated fibrils (SAFs) were detected neither in normal control brains nor in the brains of ovines with other neurodegenerative disease.

A scrapie-specific protein of about 30,000-daltons molecular weight was identified by Prusiner and his co-workers¹¹⁾. This specific protein, called a "prion protein", aggregates into rod-shaped fibrils resembling the SAFs which Merz et al. had isolated earlier⁴⁾. Antibodies to this prion protein reacts with prion rods or SAFs from scrapie. Recent study has showed that two cases one each in Hokkaido and Kanagawa⁸⁾ were associated with a prion protein from the brain and spleen samples stocked in -20°C.

Strong resistance to usual decontamination procedure was shown in the unconventional virus causing scrapie. Autoclaving at 132°C for 1 hr and the use of 5% hypochlorite or 0.1N sodium hydroxide are recommended for disinfecting instruments and contaminated areas¹⁾. The infectious nature of scrapie was re-discovered during the 1930s in England by the serious vaccine accident, in which the formalized louping-ill virus vaccine produced using scrapie-infected sheep brain caused scrapie infection in 18,000 British sheep. Recently, a scrapie-like disease in cattle (bovine spongiform encephalopathy, BSE) has also been reported in England as a newly-recognized member of transmissible degenerative encephalopathies, of which scrapie in sheep and Creutzfeldt-Jacob disease in human beings are well known¹⁴⁾. And it has been suggested that BSE be caused by scrapie-contaminated animal protein contained in commercial cattle feed⁶⁾. The development of effective decontamination and disinfection procedure is a matter of urgency in Japan.

The authors wish to express great appreciation to Drs. Eiichi Mototani and Tomoo Yoshino, National Institute of Animal Health, for their cooperation in collecting and analyzing the data from various parts of the country.

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(Received for publication, March 19, 1990)