

P.7.- First Belgian Nor98 scrapie case diagnosed via active surveillance.

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Scrapie is a fatal transmissible spongiform encephalopathy caused by prions. Typical features of these diseases are a long incubation period and the gradual vacuolation of brain neurons and neuropil. Several (classical) scrapie strains have been described based on lesion profiling in mice. In Belgium, since April 2002, all sheep older than 18 months are tested with a rapid test (Bio-Rad) through the active TSE surveillance program (EC regulation 999/2001). Five of the 6 outbreaks in 2002 showed a classical scrapie lesion profile, but in one case the positive sheep showed special features. The ewe was apparently healthy and presented for slaughter. According to the active epidemio-surveillance protocol only part of the medulla oblongata around the region of the obex is taken out. The sample was repeatedly tested positive with the rapid test. The histopathological investigation revealed no vacuolar lesions neither in neurons and in neuropil in the region of the obex. There was no detectable PrPsc as revealed by immunohistochemistry in the obex region and tonsils. The detection of scrapie associated fibrils (SAFs) was also negative. PrPsc Western blot (WB) analysis was positive showing a PrPsc glycoprofile with a strongly marked lower band at ~ 12 kDa, compared to a classical scrapie glycoprofile. The “special” glycoprofile of the present case was confirmed by Benestad S. and coworkers in Norway. According to the surveillance protocol, the whole flock was culled and the brains of all animals older than 18 months were examined, but no other animal of the flock tested positive with the rapid test. The sheep PrP genotype was A136R154Q171 homozygous, analyzed via denaturizing gradient gel electrophoresis.

The unusual characteristics of the present case are: 1/ only one (of the 55) animals of the flock was affected; 2/ no lesions were present in the brainstem (obex) as compared to the described lesion profiles of classical scrapie cases; 3/ the absence of PrPsc immunolabelling in the area of the obex; 4/ the PrPsc glycoprofile of the present case differed clearly from the glycoprofiles found in isolates of classical scrapie strains and the BSE strain, and is not distinguishable from the Nor98 glycoprofile. All these features corresponded very well with those reported of the unusual Nor98 strain detected in Norway. This type of scrapie, described for the first time outside Norway, may question the scrapie active epidemio-surveillance protocol because: from a diagnostic point of view, the positive results obtained with a rapid test require to be confirmed by standard methods like the histopathological examination and the immunohistochemical detection of PrPsc at the level of the obex. In the present case the official Belgian recognized confirmation tests (histology, immunohistochemistry and SAF) were all negative. Only the WB confirmed the scrapie positivity of the present case and the diagnosis of scrapie could therefore have been overlooked. This may be of significance for future sampling in scrapie surveillance programs and confirmation tests.

Keywords

Sheep, NOR98, scrapie, TSE