

P.6.- Atypical case of bovine spongiform encephalopathy in an East-Flemish cow in Belgium.

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Bovine spongiform encephalopathy (BSE) is a prion disease with a fatal neurodegenerative pathogenesis. It is characterized by the accumulation of an abnormal protein (PrPres), formed posttranslationally from the normal isoform (PrPc). Research in mice showed the existence of different sheep scrapie strains. Scrapie strain discrimination is currently based upon biological typing in a panel of inbred mice. However, no large scale studies of the molecular features of PrPres have been reported for bovine BSE to date. Till now, the BSE strain seemed to maintain constant biological and molecular properties even after experimental or accidental passages into different species. Very recently, variant forms of BSE have been reported in Japan, Italy and France. These forms were characterized by atypical histopathological, immunohistochemical and/or biochemical phenotype compared to the classical BSE strain. The present case describes the first Belgian atypical BSE case. Since January 1st 2001, all cattle older than 30 months is tested for TSE via a rapid test following EC regulation 999/2001. Samples positive according to the ELISA screening are further subjected to scrapie associated fibrils (SAF), histopathology, immunohistochemistry and Western blot (WB) at the NRL. A positive ELISA sample from a 64 month-old East-Flemish or Belgian white and red cow was presented at the NRL for confirmation. The histopathology of the obex, pons and midbrain was negative, immunohistochemistry and SAF were also negative. However, WB analysis was positive with an electrophoretic profile different from that of a typical BSE case. The band on the gel of the non-glycosylated form of PrPres of the present case clearly showed a lower migratrion pattern compared to that of a typical BSE case. For many years it was assumed that there was only one BSE strain. Only very recently, reports of atypical BSE cases were announced in Japan, Italy and France. The Japanese case describes a very young bull (23 months) negative on histopathology and immunohistochemistry and a WB electrophoretic profile different from that of classical BSE. The Italians observed two BSE affected cattle with a a different staining pattern on immunohistochemistry, a difference in size and glycoform ratio of PrPres on WB and a difference in regional distribution of lesions. The French two cases showed variant molecular features with a different electrophoretic profile from other BSE cases. The present case shows the most similarities with the Japanese case (except for the age). The fact that these strains were detected worldwide and in several breeds suggest that there is no local or breed dependent feature involved. It could be that the WB techniques have become more specifique within the last year or infection of cattle by scrapie could also be considered. In conclusion, continued research on BSE reveals nowadays different BSE strains in analogy with the different sheep scrapie strains. Atypical BSE cases may question the significance and efficiency of the BSE epidemio-surveillance protocol and the validation of the confirmatory tests.

Keywords

Bovine spongiform encephalopathy, BSE, Western Blot, atypical BSE