

UNUSUAL BRAIN DISEASE DEATH IN THE UNITED KINGDOM: POSSIBLE IMPLICATIONS FOR VARIANT CREUTZFELDT-JACOB DISEASE

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A British woman who died of a brain disease with similarities to variant Creutzfeldt-Jacob disease (vCJD) in 2000 had a different genetic marker from all previous vCJD patients, according to researchers from the National Hospital for Neurology and Neurosurgery in the United Kingdom (UK) in a case report published in Archives of Neurology [1].

Cases of vCJD were first seen in the UK in 1996 following an epidemic of bovine spongiform encephalopathy (BSE) in cattle, sparking fears of a possible pandemic. The European Commission's Scientific Steering Committee - specially convened as a result of such fears - stated in 1999 that as many as 500,000 people could have been exposed to BSE from each single infected bovine, leading to speculation that millions of people could be at risk [2]. Such estimations have since appeared overly pessimistic; according to the European and Allied Countries Collaborative Study Group of CJD, 163 people have died of confirmed or probable vCJD in the UK to date [3].

Until the case described in this latest paper, all vCJD patients tested were homozygous for methionine (MM) at codon 129 of the prion protein gene (PRNP). This patient had the valine-homozygous (VV) form of the prion protein (PrP), that was previously believed to confer protection against vCJD.

The scrapie isoform of PrP (PrP^{Sc}) isolated from her brain tissue was similar to the molecular strain (PrP^{Sc} type 4) typical for vCJD, but showed an altered protease cleavage pattern in the presence of a metal chelator. This therefore raises the possibility of a new form of the disease. However, the article's authors stressed that such a conclusion cannot be drawn from one case alone.

References

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