MedicalXpress

Researchers find new piece of BSE puzzle

November 20 2009

A new treatment route for bovine spongiform encephalopathy (BSE) and its human form Creutzfeldt Jakob disease (CJD) could be a step closer based on new results from scientists at the University of Leeds. The team has found that a protein called Glypican-1 plays a key role in the development of BSE. Details are published November 20 in the open-access journal *PLoS Pathogens*.

BSE, commonly known as mad cow disease, is known to be caused by an infectious and abnormal form of the prion protein that is present on cells within the nervous system. But scientists have been unclear as to what causes the abnormality to occur.

The new research from Leeds' Faculty of Biological Sciences provides part of the answer. The researchers have shown that the presence of Glypican-1 causes the numbers of abnormal prion proteins to rise. In experiments, when levels of Glypican-1 were reduced in infected cells, the levels of the abnormal prion reduced as well.

The discovery was a mixture of scientific detective work and luck, according to Professor of Biochemistry, Nigel Hooper.

"We were looking at how the normal prion protein functions in cells and spotted that it was interacting with something," he said. "Some lateral thinking and deduction led us to Glypican-1 and when we carried out the experiment, we found we were right."

The researchers suggest that Glypican-1 acts as a scaffold bringing the

two forms of the prion protein together and that this contact causes normal prions to mutate into the infectious form. They are currently seeking further funding to investigate their hypothesis.

The findings have implications for the treatment of both BSE and the human form of the disease, CJD, according to Professor Hooper.

"Now that we know the identity of one of the key molecules in the disease process, we may in the future be able to design drugs that target this."

Although the scientists mainly conducted experiments using cells infected with prions, it is also possible that Glypican-1 is involved in other diseases of the nervous system.

"While initial experiments haven't shown any link with other neurodegenerative diseases like Alzheimer's, we're not yet completely ruling that out," said Professor Hooper.

More information: Taylor DR, Whitehouse IJ, Hooper NM (2009) Glypican-1 Mediates Both Prion Protein Lipid Raft Association and Disease Isoform Formation. *PLoS Pathog* 5(11): e1000666. doi:10.1371/journal.ppat.1000666

The authors have declared that no competing interests exist.

Source: Public Library of Science (news : web)