EDITORIAL

BOVINE SPONGIFORM ENCEPHALOPATHY AND CREUTZFELDT-JAKOB DISEASE - A BEEFY PROBLEM?

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The recent scare of the possible effects of bovine spongiform encephalopathy (BSE, mad cow disease) on human health has reached a mass hysteria. As always, mass hysteria is an epidemic, is contagious, and is based on lack of knowledge. The first epidemic, of course, was of BSE itself. This was first described in the United Kingdom in the mid 1980's and although it failed to attract attention immediately, as the numbers of affected animals rose drastically, serious investigations started and, using epidemiological techniques, have led to the tentative conclusion that BSE resulted from a change in the practice of feeding British cattle – rather than grazing in the fields, they were confined to their stalls and provided protein-rich food additives, largely made of sheep and cow offal. Scrapie, a related disease, has been prevalent among sheep and goats in England for centuries, and it was speculated, although without scientific evidence, that contamination of the food chain led to the transmission of the disease across the species barriers. Because of the high prevalence of scrapie, some material originating in sheep found its way into the bovine food chain. In 1988-9, stiff measures were taken to ensure that the procedure is stopped and the number of new cases of BSE fell dramatically. It must be stressed that although it seems logical to assume that BSE resulted from scrapie (which is naturally transmitted through feeding), there is no direct proof in this regard and doubts remain.

Speculations of a possible transmission to humans came early. The human counterpart of BSE and scrapie is Creutzfeldt Jakob disease (CJD), which shares many clinical and pathological features with the other spongiform encephalopathies[1]. CJD is rare, with an incidence of about 1 new case annually for each 1,000,000 population. These numbers have been stable, and not different in various countries, for a long time. But with the awareness of the BSE epidemic in the UK, each new case of CJD has led to a full-fledged investigation of a possible link to meat consumption. Based on available knowledge, the danger seemed slim. Scrapie does not transmit to humans, CJD is as prevalent in countries without scrapie as it is in those with a high prevalence, and a species barrier seems to prevent scrapie from spreading to humans. Therefore, why should BSE be able to cross to humans? And, indeed, for several years the rate of CJD in the UK remained low. Although several individual CJD cases made it to the media, who were quick to report if any of them was somehow connected to livestock or even visited a farm, there was nothing to justify alarm. If a slight increase in incidence was noted, it could easily be explained as being due to greater awareness. In fact, a registry of CJD, established in the United Kingdom several years ago by Professor WB Matthew, was a cornerstone to this surveillance.

The recent report by Will et al[2] of 10 cases of CJD occurring in young people in the UK changed the perspective overnight. These workers reported on 10 new cases of CJD which have occurred over about one year. Although the effect on the general incidence figures was insignificant, these new cases were remarkable because of their young age (15-40 at onset), somewhat unusual course, and the fact that the pathology, although undoubtedly of a spongiform encephalopathy, was unlike that which had been previously recognised. The uniqueness of these cases led the researchers to suggest, although very carefully, at the possible link to BSE.

Several reservations are indeed appropriate. The cases reported by Will et al[2] were younger than usually seen previously, but it is not impossible that the pathology and clinical manifestations of the usual form of CJD are age-dependent, rather than necessarily related to a bovine source. A similar case has now been reported from France[3] where BSE is extremely rare. The clinical heterogeneity of human spongiform encephalopathy is still unfolding[4] and new aspects of the pathology are also being reported. So more data will be needed before a firmer conclusion can be drawn.

Some data may come from transgenic animals. Mice carrying the human form of the PRNP gene were created[5] and therefore the theory that BSE can be transmitted to humans more easily than scrapie - perhaps because normal human PRNP is more similar to that of the cow than of the sheep – can be tested. However, these studies will take time, and will not necessarily provide the final answer. It is likely that in the meantime epidemiological data, provided by goods surveillance, will be available to determine whether indeed the number of CJD cases in the UK has been increasing.

The simplistic hypothesis that CJD results from eating cattle products also has to explain why the new form occurs just in young people. Perhaps the reason is that they are the ones who crowd at fast-food chains. If this is the reason, it will strengthen the hypothesis that it is not beef meat but perhaps brain (used in making hamburgers, sausages etc), which carries the infective material.

One of the characteristics of spongiform encephalopathies is the long incubation time from inoculation to clinical manifestations, usually of several years. If the new cases which have now appeared are due to BSE, most likely the exposure occurred in the 80's. The first cases of BSE were diagnosed around 1985, and with an incubation time of 2 or 3 years, the earliest possible exposure was in 1982 (the change of the feeding pattern had occurred in 1980). With all the unknowns, particularly the length of the incubation period and the infectivity of food made of BSE-affected animals, the possibility exists that a CJD epidemic has begun in England (and may also affect those who visited there during the 1980's). On the more optimistic side, if those young people who came down with CJD were exposed in

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1989 - at the height of the BSE epidemic - the situation is a lot less ominous.

Spongiform encephalopathies, in animals and humans, are very unusual diseases. Much research was invested in studying the disease mechanisms, and much was gained. Unfortunately, few attempts addressed themselves to curing the disease, and it is a matter of urgency that we do so now, using animal models which were developed over the past few years.

References

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