LETTERS TO THE EDITOR

Familial Pick’s disease and dementia in frontal lobe degeneration of non-Alzheimer type are not variants of prion disease

The prion diseases are a group of neurodegenerative conditions affecting both humans and animals. They are transmissible after inoculation, have long incubation periods, and have been known as the spongiform encephalopathies, slow virus diseases, or transmissible spongiform encephalopathy (TSE). The prion protein (PrP) is an abundant component of the brain tissue that, in the absence of any detectable abnormality, can be transmitted to other species when it is inoculated into them. Prion diseases are caused by the accumulation of an abnormal form of the prion protein, which is thought to be derived from the normal form.

Familial Pick’s disease (FPD) is a neurodegenerative disorder characterized by the presence of neurofibrillary tangles in the cerebral cortex, particularly in the frontal lobes. These tangles are composed of hyperphosphorylated tau protein, which forms paired helical filaments. The clinical features of FPD include progressive cognitive decline, motor dysfunction, and behavioral changes.

Dementia is a general term for a syndrome characterized by the loss of cognitive function severe enough to interfere with social or occupational functions. It is a common condition in older adults and is associated with a variety of underlying causes, including neurodegenerative diseases such as Alzheimer’s disease and frontotemporal lobar degeneration.

The presence of both classical clinical and histological criteria in which the disease segregates shows an autosomal dominant pattern. All families have been documented previously.1-6

We would like to bring to your attention the possibility that the diseases described here may represent a new class of prion diseases. The clinical and pathological features of these conditions are similar to those seen in prion diseases, but the genetic basis is not yet clear. Further research is needed to fully understand the nature of these disorders and their potential role in the development of prion diseases.


Acute neuromuscular respiratory paralysis

In their article on respiratory failure Hughes and Bhatia8 made the point that forced vital capacity (FVC) and not peak expiratory flow (PEF) should be used in the assessment of declining respiratory function. The FVC has been shown to have predictive value for the need for ventilatory assistance in patients with myasthenia gravis, but the results of a survey including the non-availability of such facilities. A 65-year-old woman with myasthenia gravis, affecting mainly eye movement, bulbar function, facial expression, and neck strength was treated with increased dose of pyridostigmine and later started on 75 mg prednisolone daily. As may have been predicted, after six days of the steroid her clinical condition worsened. This point serial measurements of both FVC and PEF were measured (figure). An increased dose of pyridostigmine was prescribed and neostigmine added. During the