RAPID NEUROLOGICAL DECLINE AND DEATH IN A 79-YEAR-OLD WOMAN

To the Editor:

In March 2003, a 79-year-old woman was referred to our hospital for evaluation of recurrent falls, lumbar pain, and probable depression. She had a history of unarticulated speech, concentration, and increasing memory problems that began in 2002. In February 2003, her family physician noticed physical and mental decline and a tremor of the arms and legs. The patient fell repeatedly and was unable to walk without help. On admission, the patient herself complained of fatigue. Her past medical history was positive for hypertension, peripheral arterial vascular disease with femoral bypass operation, and carpal tunnel syndrome. Her review of systems revealed dysuria. She presented mentally slowed but well oriented. Physical examination revealed a low frequency tremor of her arms and legs, decreased coordination, weak reflexes, and a reduction of her vibratory senses. She was unable to walk due to gait ataxia. Laboratory results were within normal range and her urinalysis revealed a bladder infection, which was treated with antibiotics. The patient was currently taking coumadine, amlodipine, rofecoxib, acetaminophen, and a tranquilizer. To rule out a chronic subdural hematoma, the patient had an emergency CT brain scan, which showed old diffuse ischemic white matter changes. Within 2 days, she developed progressive ataxia, choreatic movements of both arms, dysarthria, and dysphagia. At this time, she was no longer able to participate in a mental status exam. Consequently an MRI scan of her head and vertebral spine was ordered. The spine scan revealed a fracture of vertebra T8. The brain scan demonstrated bilateral cortical signal enhancements in the T2-weighted sequences. These lesions were highly suspect of sporadic Creutzfeldt-Jakob disease (CJD). Further examinations such as EEG and a lumbar puncture were not performed due to the patient’s rapid deterioration. In addition, the patient’s family members declined to consent. During the subsequent days, the patient developed anarthria, progressive tetraparesis, hyporeflexia, and myocloni. She lapsed into a coma and died 12 days after admission. Histopathology of her brain assessed by the Swiss National Reference Center for Prion Diseases revealed gliosis, astrocytosis, and diffuse spongiform changes of the entire cortex, the basal ganglia, the brain stem, and the cerebellum, respectively. Immunohistochemistry stains were positive for prion protein within the cortex and the basal ganglia, a finding that was confirmed by Western blot. Thus, the patient died of sporadic CJD.

Comment

Although sporadic CJD is a rare disease with an incidence of 1/million/year and an average age of onset between 50 and 70 years (1), it should be considered in aged persons with neurological and/or cognitive decline. In contrast to an earlier report in a somewhat younger patient (2), cognitive function in our patient declined slowly and was initially misinterpreted as depression or dementia of another type, such as Alzheimer’s disease. Finally, the patient’s family members declining consent for diagnostic procedures is a common problem in geriatric medicine in Europe. However, a diagnosis of CJD is also possible through a history and an MRI examination, which is a reliable noninvasive diagnostic tool (3).

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