Letter to Editor

A rare case of sporadic Creutzfeldt–Jakob disease in Isfahan, Iran

To the Editor
We would like to report a rare case of sporadic Creutzfeldt-Jacob disease in Isfahan, Iran. A 61-year-old man without familial or personal history of neurologic disease or hormone treatment presented initially with problems of comprehension and psychomotor condition.

On admission to the neurology department, in Kashani hospital, Isfahan, about three months after the first symptoms, neuropsychological examination showed drowsiness, apathetic affect, decreased speech output, visual hallucination, negative attitude, concrete thinking, and impaired judgment. Neurologic examination showed augmentation of muscle tone (rigidity), spontaneous myoclonus involving entire body, impaired pursuit gaze, asterixis and ataxic gait. Vital signs were normal. There were no abnormalities on extensive laboratory and radiological screening, including liver function tests, blood culture, serum HIV antibody tests, serum and CSF HSV–PCR, CSF cytology, thyroid survey, serum Vitamin B12 and lithium levels, CSF VDRL, CSF India ink test. The EEG showed one per second periodic paroxysm of sharp waves on a slow background. In conventional sagittal and transverse spine echo MRI, no significant visible atrophy or signal change was present. In neuropathologic investigation, two brain samples (open biopsy) from fronto–parietal regions were taken. Hematoxylin and eosin (H & E) staining showed non-inflammatory vacuolization of the cerebral cortex with neuronal atrophy and astrogliosis. The density of astrocytic gliosis was analyzed on H & E and on immunostained section using anti-glial fibrillary acidic protein antibody. The patient deteriorated progressively and developed a kinetic mutism after 15 days. Death occurred about 45 days after admission and 4.5 months after the appearance of the first symptoms.

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