A Rapidly Progressive Dementia: Think Creutzfeldt-Jakob Disease

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Introduction

Creutzfeldt-Jakob Disease (CJD) is the most frequently occurring human prion disease. The annual incidence rate is 1-2 per million worldwide. The vast majority of cases occur as a sporadic disorder. We present a 71-year-old man with a history of cognitive decline, myoclonus, cerebellar signs and aphasia. Neuroimaging and CSF analysis supported a diagnosis of probable sporadic CJD (sCJD).

Description of Case

This 71-year-old man initially presented with a 3-month history of gait unsteadiness and increasing confusion. On examination he had an ataxic gait. Mini-Addenbrooke's Cognitive Examination (Mini-ACE) score was 14/30. Initial MRI brain showed no significant abnormality.

Over the following 2 months his symptoms rapidly progressed. His mobility deteriorated, he developed myoclonus and became progressively more aphasic. Repeat MRI brain showed bilateral diffusion restriction involving the caudate nuclei, putamini and thalami. EEG showed periodic sharp wave complexes. Cerebrospinal fluid was positive for Real-time quaking-induced conversion (RT-QuIC). Therefore our patient met the criteria for a diagnosis of probable sCJD (as per Euro-CJD classification). He continued to decline and died 6 months from symptom-onset.

Discussion

sCJD is rare however should always be considered in a patient presenting with a rapidly progressive dementia, particularly if accompanied by myoclonus and/or cerebellar signs.