Ana Ivancheva et al., J Alzheimers Dis Parkinsonism 2017, 7:4(Suppl)
DOI: 10.4172/2161-0460-C1-028

Prion disease – The last diagnosis

Ana Ivancheva and **Hakim Ghani**
Mayo University Hospital, Ireland

A 73-year-old lady was referred to a neurologist a er two geriatric consults due to deteriorating cognitive function, ataxia and weakness. She was completely dependent in slightly over one year. From walking with a frame, she became wheelchair bound. Her cognition continued to decline and could not feed herself due to apraxia. When assessed she was found to have rigidity, myoclonus and le -right disorientation. Overall impression was that she had prominent extrapyramidal and cortical disease.

Diagnosis: Due to rapid symptoms progression, typical MRI brain ndings and highly speci c CSF 14-3-3, S100B and RT-QuIC, she was diagnosed with probable sporadic Creutzfeldt-Jakob disease(sCJD). Brain biopsy post-mortem would con rm the diagnosis.

Discussion: SCJD has unclear mechanism of initial conformational change of prion protein. CJD has worldwide distribution, where 85% are sCJD and occurs at rate of 1 in 1,000,000 population per year. Mortality rate is less than 1 per million depending on country. Researchers mention di culties in ascertaining accurate incidence, prevalence and mortality rate due to under-reporting, under-awareness and poor diagnostic capability. Possibly CJD is higher than thought due to misdiagnosis

Biography

Notes: