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Clinical Image

Case of Sporadic Creutzfeldt Jacob Disease

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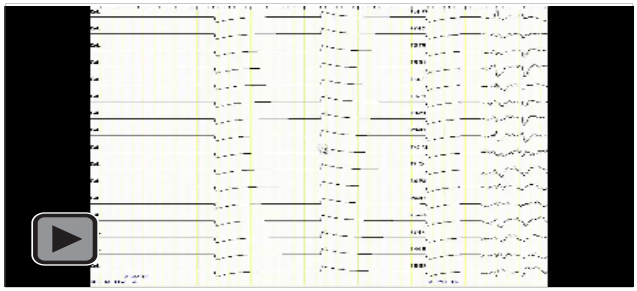
Clinical Image

A 67 year-old man presented with a two-month history of memory loss and speech difficulties. His family first noted behavioral changes and signs of depression, a few months earlier. The symptoms had deteriorated rapidly during the previous three weeks rendering him incapable of performing everyday activities. Mental status assessment revealed severe impairment of all cognitive functions and language skills with prominent verbal perseveration. The neurological examination showed signs of ideomotor apraxia, appendicular ataxia and asymmetric, irregular myoclonus of the upper limbs (Video 1). Brain MRI was unrevealing. An EEG was performed demonstrating generalized periodic triphasic sharp wave complexes (1-2/sec) characteristic of sporadic Creutzfeldt-Jacob disease (Video 2). CSF analysis showed mildly increased protein as well as an elevated 14-3-3 protein level. During the next two months, the symptoms took an unrelenting course progressing to a state of akinetic mutism and the patient died approximately 6 months after onset.

Video 1:



Video 2:

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