Abstract

Background

Creutzfeld Jacob disease (CJD) is a rare transmissible neurodegenerative disorder with relentless progression,rapid neurocognitive deterioration and invariable death. In its early presentation it can closely mimic other forms of slowly progressive neurodegenerative disorders without a significant cognitive impairment. Alternatively in its phase of rapid cognitive decline it could be mimicked by many treatable conditions such as autoimmune/paraneoplastic encephalopathy in the elderly population. Hence the differentiation of this universally fatal condition is of greatest therapeutic significance.

Case

This is a case of a 60year old previously well male presenting with cerebellar ataxia, rigidity, erectile dysfunction and significant postural blood pressure drop subsequently developing rapidly progressive dementia with prominent visual symptoms and stimulus sensitive action myoclonus.

Conclusion

Sporadic Creutzfeld Jacob disease presents with prominent cerebellar ataxia in its early phase.