

Abstract

Background

Creutzfeldt-Jakob disease (CJD) is the most common of the prion diseases which can cause rapidly progressive dementia. In addition it can have other features such as psychiatric manifestations, myoclonus etc. We report the case of a 74 year old Sri Lankan lady who presented with rapidly progressive dementia and was diagnosed to have sporadic CJD.

Case Presentation

74 year old Sri Lankan female diagnosed patient with hypertension and dyslipidaemia presented with rapidly progressive dementia and abnormal behaviour for 2 months duration and walking difficulty and mutism for one month duration. On examination she was found to be rigid with right upper limb myoclonus. Her basic haematological investigations and inflammatory markers were normal. The MRI of the brain was suggestive of characteristic cortical ribboning pattern of sporadic CJD. CSF full report was normal. EEG demonstrated the characteristic periodic sharp wave complex. The patient was diagnosed to have sporadic CJD.

Conclusion

It is important to look for an underlying cause in patients with rapidly progressive dementia since some of the underlying conditions are potentially treatable. Even though it is a rare diagnosis in Sri Lanka it is important to look for prion diseases in patient with rapid onset dementia and movement disorders.