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PROBABLE CREUTZFELD-JAKOB DISEASE: A NEUROPSYCHIATRIC PRESENTATION

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Creutzfeld-Jakob disease (CJD) is the commonest form of transmissible human subacute spongiform encephalopathy, with a worldwide distribution of 0.5 to 1.0 per million population. A retrospective review in 1998 from Singapore General Hospital revealed 5 possible cases over 2 years, in an island with a population of 4 million. We present a 76 year-old Chinese man, with a premorbid personality of an independent person who was neat and meticulous, admitted for a 3 month-history of progressive behavioural change, associated with memory impairment and unsteady gait. He had long-standing hypertension which was well-controlled. The abnormal behaviour included frequent complaints of people coming to his house to have meetings, frequent misplacing of his belongings, as well as decreased self-hygiene. This has affected his daily functioning. There were no other symptoms of dementia. Initial MMSE was 16/30, with impairment in 2-point orientation (place/person), the serial sevens, and delayed recall. There was marked inattention. There was no evidence of mood disorder. No perceptual disorder was elicited. The brain MRI revealed cortical hyperintensities ("cortical ribboning") in right frontal, parietal, temporal, and caudate nucleus. Physical examination on the 5th day of admission revealed mild bradykinesia, mild hypertonia of all 4 limbs, mildly impaired proprioception, mildly unsteady gait, as well as occasional myoclonic jerks of the head. Electroencephalogram showed generalised fronto-central fairly rhythmical periodic complexes with triphasic morphology.Cerebrospinal fluid protein 14-3-3 were negative. Over the next 12 days, his Abbreviated Mental Test (AMT) decreased from 8/10 to 5/10. The clinical picture was highly suggestive of sporadic CJD.

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