

Cortical Ribboning and Striatal Diffusion Restriction in Suspected Creutzfeldt–Jakob Disease

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Abstract:

Creutzfeldt–Jakob disease (CJD) is a rare, rapidly progressive neurodegenerative disorder characterised by cognitive decline, behavioural abnormalities, myoclonus, and pyramidal or extrapyramidal signs. Magnetic resonance imaging (MRI), particularly diffusion-weighted imaging (DWI), plays an important role in its diagnostic evaluation. A 69-year-old man presented with a 5–6-month history of progressive behavioural disturbances, severe memory impairment, disorientation, and executive dysfunction, accompanied by stimulus-sensitive myoclonic jerks. Neurological examination revealed inattention, spasticity involving all four limbs, symmetrical mild quadriparesis, brisk deep tendon reflexes, a positive applause sign, and impaired motor sequencing. Routine investigations, including thyroid function, renal and liver function, serum vitamin B12, and viral markers, were unremarkable. Plain MRI of the brain demonstrated T2/FLAIR hyperintensity with corresponding diffusion restriction involving the cortical ribbon of both cerebral hemispheres, sparing the left occipital lobe and bilateral precentral and postcentral gyri. Additional areas of restricted diffusion with corresponding FLAIR hyperintensity involved the heads of both caudate nuclei and bilateral putamina, with greater involvement on the right. The combination of cortical ribboning and striatal involvement, together with rapidly progressive cognitive decline and myoclonus, raised strong suspicion of CJD. This case highlights the diagnostic value of recognising characteristic diffusion abnormalities in patients presenting with rapidly progressive dementia. Although these MRI findings support the diagnosis, they are not independently definitive; correlation with electroencephalography, cerebrospinal fluid real-time quaking-induced conversion testing, and exclusion of relevant mimics is essential. Early recognition of this imaging pattern can facilitate appropriate diagnostic evaluation and clinical counselling.

Keywords:

Creutzfeldt–jakob disease, cortical ribboning, diffusion-weighted imaging, caudate nucleus, rapidly progressive dementia.