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123I-ioflupane SPECT scan in a patient with Creutzfeldt-Jakob Disease

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Key words: Creutzfeldt-Jakob; single photon emission computed tomography; SPECT; 123I-FP-CIT; ioflupane.

Case summary

A 62-year-old woman presented with gait ataxia, bradykinesia, bilateral rigidity, choreic and dystonic movements, and psychomotor slowing. Magnetic resonance imaging (MRI) (Fig. 1), EEG and CSF analysis were compatible with a diagnosis of Creutzfeldt-Jakob disease (CJD). The 123I-ioflupane SPECT (Fig. 2) was normal, indicating the relative sparing of nigro-striatal fibers and suggesting that her movement disorder was due to postsynaptic dysfunction. Post-mortem examination was not performed. MRI findings in CJD are well known, showing typically hyperintense signals in striatum and cortex on T2-weighted, fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted images. But there is no correlation between hyperintense basal ganglia on MRI and extrapyramidal signs (Meissner *et al.*, 2004).

Surprisingly, in vivo measures of neurotransmission in CJD are very rare. We found only three 123I-ioflupane SPECT reports in the literature. Two of them – a "parkinsonism" (Lipczynska-Lojkowska *et al.*, 2007), and a corticobasal syndrome (Vandenberghe *et al.*, 2007) – were normal. The third

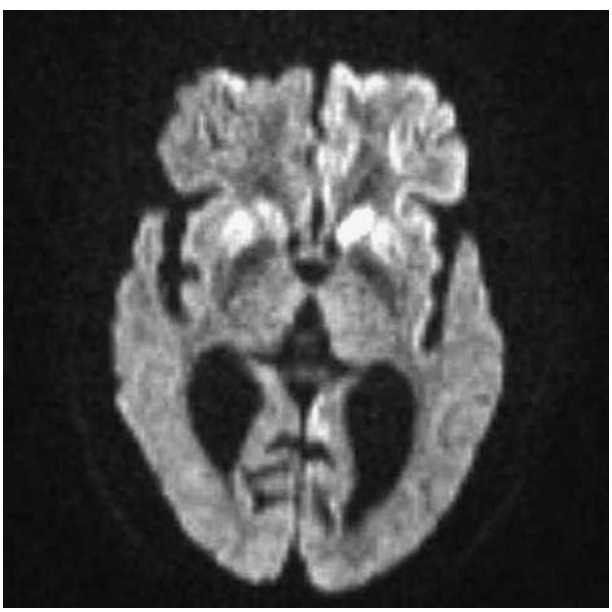

Fig. 1. — Brain MRI (diffusion-weighted image): hyperintense area in putamen, caudate nuclei, and left frontal, frontobasal, and external temporal cortex.

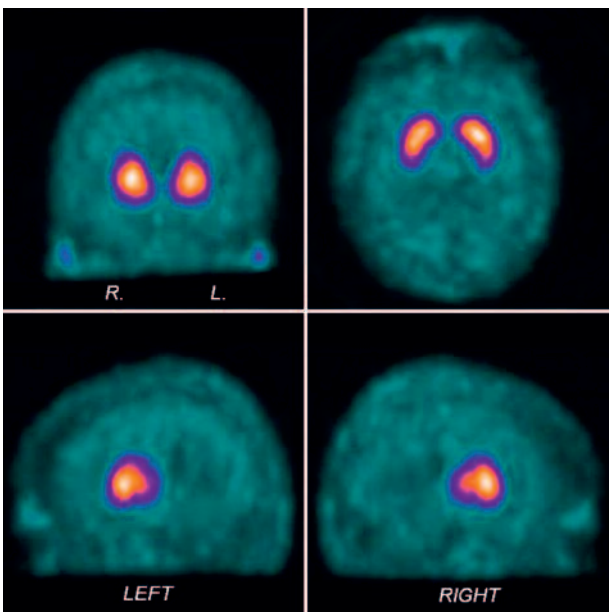

Fig. 2. — 123I-ioflupane SPECT scan (six weeks after the onset of the symptoms): normal.

one (a patient with cognitive decline and right hemiparesis and extrapyramidal rigidity) disclosed reduced value of the radiotracer in the left putamen (Ragno *et al.*, 2009). Single cases studies may result in heterogeneous findings due to phenotypic differences, or time between first symptoms and SPECT (3 weeks in our case, 3 months in (Ragno *et al.*, 2009)). Therefore larger groups are needed. These might disclose neural transmission abnormalities, which remain widely unknown in this disease.

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