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

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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 (Vandenberghhe et al., 2007) – were normal. The third

Fig. 1. — Brain MRI (diffusion-weighted image): hyperintense area in putamen, caudate nuclei, and left frontal, fronto-basal, 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.

REFERENCES


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