

death, she developed subacute auditory hallucinations and persecutory delusions (for example, she thought someone was intruding into and wiretapping her house, or believed that her child had been kidnapped). A few days later, she became excitable and tried to strangle her husband, after which she was restrained by her family and admitted to our hospital. On admission, she was alert, and physical and neurological examinations revealed no abnormalities. Laboratory tests were normal. A magnetic resonance imaging (MRI) scan revealed a subacute right putaminal infarction, which was partially extended to the right caudate and insula. The infarction was treated conservatively. Administration of risperidone (5mg/day) was initiated and the psychotic symptoms gradually disappeared. Two months later, the psychotic symptoms had completely disappeared and she was discharged. The regimen of risperidone was tapered off over the following 6 months and no relapse was observed.

#### Comment

Cummings<sup>1</sup> noted that subcortical structures (striatum [caudate and putamen], globus pallidus, and thalamus) and the prefrontal cortex form three circuits (dorsolateral prefrontal, lateral orbital, and anterior cingulate circuits) and these circuits mediate many aspects of human behavior. On the other hand, recent neuroimaging studies in schizophrenia have demonstrated morphological and functional abnormalities of the frontal-subcortical circuits, although these results remain ambiguous.<sup>2,3</sup>

In our patient, a right putaminal infarction, which affects the dorsolateral prefrontal and lateral orbital circuits, caused schizophrenia-like hallucinations and delusions. In contrast, a case of schizophrenia in

which hallucinations and delusions disappeared following left putaminal hemorrhage was previously reported.<sup>4</sup> In this previous case, it could be speculated that excessive left frontal-subcortical function was associated with schizophrenic symptoms, and that correction of the excess function by left putaminal hemorrhage extinguished the symptoms. Collectively, these findings suggest that left and right frontal-subcortical functional asymmetry (i.e., relative right hemispheric hypofunction or relative left hemispheric hyperfunction) may play a critical role in the appearance of schizophrenic symptoms, especially for hallucinations and delusions.

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## Psychogenic Movement Disorder Masquerading as CJD

*SIR:* Creutzfeldt-Jakob Disease (CJD) is a rare, rapidly progressive prion disease characterized by dementia, dysarthria, and movement disorders, and typically culminates in death within 6 months of contraction.<sup>1</sup> We describe a patient who presented with a conversion disorder resembling CJD confounded by 14-3-3 proteins in his CSF.

#### Case Report

A 58 year-old man with a history of major depression complained of having memory impairment, difficulty walking, choppy speech, and incoordination for 1 month. There was no family history of movement disorders or dementia.

Initial examination by other physicians revealed intact immediate and recent memory and normal speech and language. He was noted to have complex choreoathetotic movements in his arms, startle myoclonus, bilateral intention tremor, and a wide-based slow gait.

Given subjective subacute memory decline, startle myoclonus and gait ataxia, the possibility of CJD was entertained. EEG and brain magnetic resonance imaging (MRI) were normal; however, 14-3-3 proteins were present in his CSF and he was diagnosed with CJD by other neurologists.

Despite complaints of new or worsening symptoms, there was no change in his examination; a repeat EEG was normal and neuropsychological testing was not consistent with a progressive dementia. His Mini-Mental State Examination

(MMSE) score was 26/30 1 year after presentation and a repeat MRI was normal 18 months after presentation. Given the long duration of nonprogressive symptoms/signs and a placebo (IV normal saline) response for most of his symptoms (administered by his primary physician), he was diagnosed with conversion disorder.

Four years later, the patient remains convinced that he has CJD, and is seeking evaluation at two other academic medical centers. His symptoms continue to fluctuate and he responds to placebo for numerous complaints.

#### Comment

The triad of ataxia, myoclonus, and progressive memory decline in a middle-aged patient should raise suspicion for CJD. The median survival after diagnosis of CJD is 4.5 months with only 10% of patients surviving for 12 months.<sup>1</sup> Our patient has survived 4 years with a nonprogressive course, making the diagnosis of CJD highly unlikely. These features, in addition to placebo-responsive symptoms/signs and comorbid depression make our

patient's illness most likely psychogenic.

CJD can present with neuropsychiatric features, including depression and even hysteria.<sup>2</sup> Unlike the initial incorrect diagnosis of CJD in our patient, most of the literature supports the underdiagnosis of CJD in this setting.<sup>3</sup> EEG and MRI can be normal in patients with CJD; however, typical findings of pseudoperiodic sharp wave complexes on EEG and hyperintensity in the basal ganglia on MRI are very helpful and specific in corroborating the diagnosis.<sup>4</sup>

While the presence of 14-3-3 protein in CSF is highly sensitive and specific for CJD in many reports, 14-3-3 proteins are found in the CSF of many patients with other neurological disorders.<sup>5</sup> Our patient did not exhibit clinical findings consistent with any other metabolic, inflammatory, infectious, vascular, or paraneoplastic causes for elevated 14-3-3 proteins in CSF.<sup>5</sup>

In patients with cognitive decline, myoclonus and ataxia, the diagnosis of CJD should be considered. Disease duration longer than 1 year and lack of a rapidly progressive

dementia are suggestive of another cause, even in the setting of elevated 14-3-3 proteins in the CSF.

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#### Correction

In the Letter, "Insulinoma in differential diagnosis of seizure disorder," in the Spring issue, the authors' names and surnames were reversed. They should have read, Dr. Murat Alemdar, Dr. Pervin Iseri, and Dr. Sezer Sener Komsuoglu.