Catatonia and Klüver-Bucy Syndrome in a Patient With Acute Disseminated Encephalomyelitis

To the Editor: Klüver-Bucy syndrome (KBS), described originally in monkeys and later in humans, is associated with bilateral anterior damage on the temporal poles, amygdala, or mediodorsal thalamic nucleus,[1,2] which are part of Yakovlev's circuit or lateral limbic circuit.[1] The uncinate fasciculus and other tracts are critical parts of this network[3]; however, it is uncertain if white matter lesions could produce KBS. We present the case of a young woman who attended the National Institute of Neurology and Neurosurgery of Mexico with acute disseminated encephalomyelitis, a clinical entity with relevant white matter damage, presenting with acute catatonia and chronic KBS.

CASE REPORT

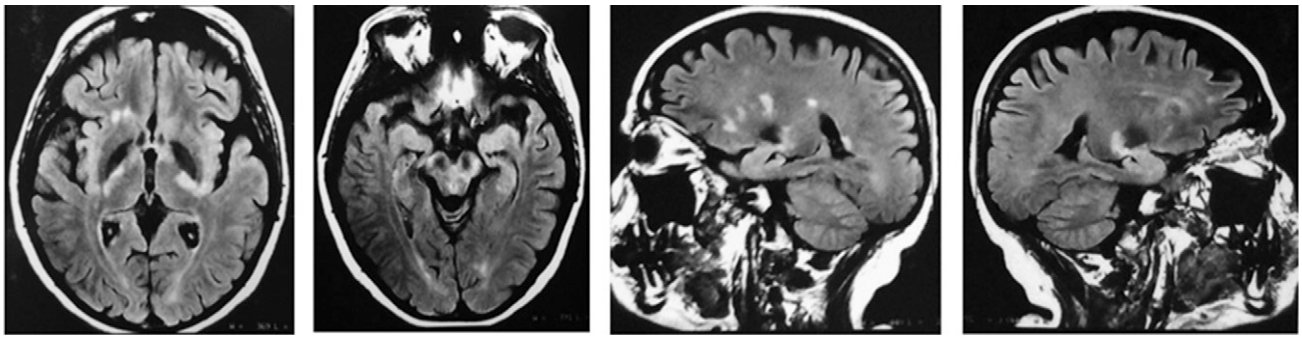
Catatonic Syndrome in a Patient With Acute Disseminated Encephalomyelitis

At age 26 years, patient "M" was a married woman, graduated as a physician, had one daughter, and did not have relevant pathological, psychiatric, or family antecedents. She received a seasonal influenza vaccine and H1N1 vaccine on November 2009. Her illness began on January 2010, with rhinorrhea, nonproductive cough, 40°C fever, headache, hypersomnia, dysarthria, and left pyramidal syndrome. Neurological examination findings at the National Institute of Neurology and Neurosurgery revealed somnolence and ophthalmoplegia, left ptosis, and left facial paralysis. Her muscle strength was 4/5 on all four extremities; generalized hyperreflexia, a dystonic posture of all four extremities, bilateral dysmetria, and indistinct lateropulsion were also documented. Laboratory testing ruled out renal, hepatic, or endocrine impairments and electrolyte imbalance. HIV, VDRL, and antinuclear antibody test results were negative. Results on adenosine deaminase, India ink microscopy, and JC virus (or John Cunningham virus) polymerase chain reaction were also negative. Results on MRI scan (February 2010) showed hyperintense lesions on T2/fluid-attenuated inversion recovery sequences on the right-sided caudate nucleus, globus pallidus, putamen, anterior thalamus, and cerebral peduncle. A second MRI scan (March 2010) showed new lesions on the contralateral side, with a similar topography. After excluding other causes, we concluded that she fulfilled criteria for acute disseminated encephalomyelitis, due to previous viral infection symptomatology; multifocal neurological involvement and encephalopathy; generalized dysfunction in the EEG; normal CSF analysis; and asymmetrical, diffuse lesions in MRI, suggestive of a demyelinating cause. Methylprednisolone pulses were followed by oral prednisone. During hospitalization, patient M developed catalepsy, mutism, bizarre postures, negativism, paratonia, gegenhalten sign, and occasional automatic obedience and grasp reflex; a catatonic syndrome was diagnosed. A rapid resolution of catatonia followed the administration of lorazepam and amantadine. She recovered alertness and was discharged with prednisone treatment.

Disruptive Turn for Patient M

Although a complete motor recovery was observed after physical therapy, insidious behavioral changes appeared 1 month after discharge. Previously described as a shy person who enjoyed academic rather than social activities, she began to approach relatives and strangers to ask them questions about their sex life. She would sneak into the men's room repeatedly to measure their genitalia and often would touch the private parts of strangers (men and women alike) and relatives (her sister and both of her parents). She began to introduce all kind of objects in her mouth to taste them and maintained food in the oral cavity for minutes or hours; dental treatment was required because she lost all of her teeth. There were important changes in dietary habits in which she began preferring candies, chocolates, and bread and avoiding water, fruits, and vegetables. She experienced severe episodes of constipation, which led to brief hospitalizations. Once, while changing her daughter's diapers, she manifested coprophagia. Her attention easily focused on new environmental stimuli, and she would ceaselessly grab surrounding objects to explore them briefly. A ludic and puerile attitude was permanent; she demanded attention from relatives, friends, and physicians. She also neglected her maternal cares and her own personal grooming. A subsequent MRI scan showed new lesions on the right orbitofrontal cortex and bilaterally on the insular lobe, the amygdala, and the white matter connecting temporolimbic structures with the ventral frontal cortex (Figure 1). A diagnosis of multiphasic acute disseminated encephalomyelitis was made by the multiple sclerosis clinic; sequential immunological treatments included methylprednisolone pulse therapy, plasmapheresis, cyclophosphamide, and azathioprine. A 4-year follow-up showed a moderate decrease in hypersexual behaviors, although patient M still runs away from time to time to have sexual intercourse with strangers. Neuropsychological testing results ruled out visual agnosia, language disorders, or significant memory disturbances. She had preserved executive functions. However, social emotional disturbances impeded her capacity to work as a physician and negatively affected her

FIGURE 1. MRI Scan Taken 5 Months After Illness Onset: Fluid-Attenuated Inversion Recovery Sequence[a]



[a](A) Bilateral hyperintensities on insulae and deep white matter hyperintensities on right orbitofrontal cortex. (B) Bilateral hyperintensities on amygdalae and entorhinal cortex. (C) Hyperintense signal on left amygdala. (D) Hyperintense signal on right amygdala.

everyday life functioning; she lives with her parents after her husband asked her for a divorce. As her insight increased, she reported depressive cognitions coexisting paradoxically with the placidity symptom of Klüver-Bucy syndrome (KBS).[4] For example, she reported feeling sad, hopeless, and alone because her life had no purpose or meaning while smiling and behaving with a childish attitude. Suicide attempts prompted treatment with electroconvulsive therapy. Sequential trials with fluoxetine, mirtazapine, trazodone, duloxetine, olanzapine, quetiapine, clozapine, aripiprazole, valproic acid, carbamazepine, methylphenidate, and cyproterone were ineffective. Cognitive-behavioral and neuropsychological rehabilitation interventions showed poor results.

DISCUSSION

To our knowledge, this case is the second to describe acute catatonia in a patient with acute disseminated encephalomyelitis.[5] Catatonic signs were possibly related to extrapyramidal damage observed in early MRI studies. Regarding KBS, patient M fulfilled five diagnostic criteria, including hypermetamorphosis, hyperorality, dietary changes, hypersexuality, and emotional and social changes (placidity, taming, anosodiaphoria, and significant reductions in the following phenomena: fear, aggression, nurture behavior, and personal space behavior regulation). The last criterion (visual agnosia) was absent. As known, a diagnosis is made with three of six symptoms.[1] Patient M also poses a problem with cognitive-behavioral dissociation. In humans, KBS is usually accompanied by extensive cognitive deficits[1,6]; despite the dramatic social-emotional disturbances, patient M developed KBS without significant impairment on memory, gnosias, praxias, executive functions, or cognitive insight. Probably, bilateral damage in multiple nodes of Yakovlev's circuit created a disconnection syndrome, leaving cortically dependent heteromodal functions relatively intact. Regarding etiology, KBS has been described in patients with herpes simplex encephalitis, Alzheimer's disease, surgical resection of brain tumors, tuberous sclerosis, and neurocysticercosis, among others.[7,8] To the best of our knowledge, there is only one case report related to acute disseminated encephalomyelitis,[9] although previous observations in multiple sclerosis[10] suggest a disconnection mechanism in patients with KBS and white matter disease.

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