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Recurrent Klazomania Responsive to Acute Plus Maintenance Electroconvulsive Therapy

To the Editor:

We present a rare case of klazomania (compulsive shouting) that was unresponsive to several trials of psychotropic medication. An initial course of electroconvulsive therapy (ECT) led to a complete resolution of symptoms. However, within 2 months, her klazomania returned, leading to a second course of ECT. This second course again led to resolution of symptoms, but required more treatments.

CASE REPORT

Ms X. was a 69-year-old woman was referred in June 2017 for consultation and consideration of ECT. The patient described a history of mild postpartum depressed mood and a somewhat anxious temperament, but no other formal psychiatric history until 2013. At that point (2013), in context of one of the patient's sons seeking divorce, she began expressing an irrational fear for the safety of her grandchildren. This escalated into a pattern of depressed/anxious mood and eventually was accompanied by tic-like utterances, in the form of frequent, sometimes nearly constant, compulsive shouting "eh" repetitively and lasting 1 to 4 seconds. The patient found the behavior socially embarrassing and limited her social contacts because of it. This compulsive shouting did not occur during sleep and occasionally remitted when the patient was less anxious. She also displayed compulsive hand biting bilaterally and developed significant discoloration and calluses on both dorsal and palmar aspects of her hands. Family members noted her to have truncal "rocking" behaviors and that she became quite ruminative about her condition.

The patient was irregularly adherent to various trials of lorazepam, clonazepam, quetiapine, risperidone, aripiprazole, amitriptyline, and sertraline (doses were not known to patient/family). These medication trials were generally ineffective except for some improvement with lorazepam and clonazepam. She denied any history of suicidal intent, but at one point, she did impulsively take 15 tablets of amitriptyline 10 mg. She was referred to individual psychotherapy but never engaged significantly. At the time of evaluation (2017), she was on amitriptyline 30 mg at bedtime. Although she denied symptoms of depression or anxiety disorder, she was notably ruminative and pessimistic about her condition. She had not had repetitive transcranial magnetic stimulation or ECT. She denied substance use. Family history included a sister with "crying fits" (of unclear significance) and several family members with depressive disorder.

Examination was unremarkable, except for repeated loud shouting, throughout interview, which she appeared to be able to contain only briefly, with effort; she also bit at her hands repeatedly. Her affect was dysphoric, but she denied depressed mood. She was fully oriented to time and place. Her short- and long-term memory was intact. Montreal Cognitive Assessment score was 25/30. Laboratory investigations, including lumbar puncture, were unremarkable.

Brain magnetic resonance imaging (without contrast) revealed no intracranial abnormality to explain etiology of symptoms. Scattered deep white matter fluid-attenuated inversion recovery signal abnormality was nonspecific but most likely represents chronic microvascular disease.

Ten electroconvulsive treatments, 3 times per week, bilateral electrode placement, energy 35 to 36 J (35%), with 0.5-millisecond pulse width, using a Thymatron System IV (Somatics LLC, Venice FL) machine led to complete resolution of symptoms. However, 3 weeks later, her symptoms recurred. A second course of 17 more bilateral treatments with same parameters eventually led to resolution of symptoms, which, however, were associated with post-ECT confusion. Given ruminative quality of depressive thoughts, she was started on sertraline up to 150 mg/d after the 14th treatment of the second course. She is currently in the continuation phase of ECT treatment, currently every 2 weeks for maintenance.

DISCUSSION

Klazomania, or uncontrolled, repetitive, compulsive shouting, apparently not explicitly related to contemporary emotional distress, is a rare phenomenon.[1] Those episodes of klazomania without neural injury or neurodegeneration can be conceptualized as a tic disorder, similar to vocal and motor tics classically seen in Tourette syndrome and similar disorders.[1,2] Electroconvulsive therapy–responsive klazomania was reported in a 74-year-old woman with comorbid depressive disorder that was unresponsive to multiple medications.[3] Pillai et al[4] reported a recent case of ECT-responsive klazomania who similarly had failed to respond to multiple psychotropic medications. Other cases in the literature reported successful abatement of persistent screaming with ECT.[5]

Our patient had experienced essentially constant klazomania for 4 years, with initial, subacute symptom onset in the context of psychosocial distress (the imminent divorce of her son) associated with likely catastrophic thinking about the implications of this event. Several antidepressant and other medication trials were largely ineffectual, and neurologic assessments yielded nonspecific findings and no useful interventions. Her initial course of 10 ECT treatments, 3 times a week, resulted in complete resolution of symptoms. However, beginning 3 weeks following the completion of the initial course of ECT, there was a recurrence of shouting and hand biting, so a second course of ECT treatments was initiated, which has led to a maintenance course of ECT treatment. As of the time of this submission, she remains in remission.

We describe a case of klazomania, a rarely encountered neuropsychiatric syndrome where ECT can be a highly effective treatment option. The mechanism by which ECT gives symptom relief in klazomania is uncertain. Psychiatrists

should consider the option for ECT in treatment-refractory klazomania cases.

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Treatment of Possible PERM Underlying Malignant Catatonia and Accompanying Psychotic Symptoms With Modified Electroconvulsive Therapy
A Case Report

To the Editors:

Progressive encephalomyelitis with rigidity and myoclonus (PERM) is a rare neurological disorder that involves rapid progressive muscle rigidity, difficulty with walking, hyperekplexia, and brainstem signs.[1] The clinical symptoms of PERM result from an impairment in gamma-aminobutyric acid (GABA) signaling and a decreased availability of GABA in the brain.[1] Few reports have described the treatment of the psychiatric symptoms of stiff-person syndrome (SPS) and PERM.

We present a case of possible PERM underlying malignant catatonia accompanied by psychiatric symptoms in a woman, who was successfully treated with modified electroconvulsive therapy (mECT). A 47-year-old woman with normal intelligence experienced difficulty with walking, and developed incontinence, incoherence, and soliloquy within a month. Psychiatrists in another hospital ascertained micromania, Capgras delusions, Cotard delusions, and visual and cenesthetic hallucinations, before she was transferred to our hospital.

The patient's vital signs were normal. She was bedridden, almost completely nonresponsive with a partial ability to follow visual cues, and was experiencing dysarthria, dysphagia, and urinary retention. The patient scored E4V2M4 (E = eye opening, V = verbal response, M = motor response) on the Glasgow Coma Scale, and had a modified Rankin Scale (mRS) score of 5. Her axial and limb muscles were markedly rigid, and she had hyperreflexia including extensor plantar reflexes. Myoclonus was absent. Sudden touch or noise elicited muscle spasms in both the axial and proximal limb muscles, indicating hyperekplexia. She exhibited increased psychotic agitation. Routine blood and cerebrospinal fluid (CSF) analyses were unremarkable. The serum anti-GAD65 antibody level was normal (<1.5 U/mL), and the serum and CSF tests for the anti-glycine receptor (GlyR) antibody and serum antineuronal antibodies were negative. Tests for antibodies against LGI1, Caspr2, NMDAR, AMPAR, $GABA_AR$, $GABA_BR$, mGluR1, and mGluR5 were also negative. Brain magnetic resonance imaging (MRI) results with gadolinium enhancement, spinal MRI at the cervicothoracic level, electroencephalography, nerve conduction study, and whole-body computed tomography with enhancement were unremarkable. Needle electromyography of the biceps and vastus medialis revealed continuous motor unit activity at rest that was completely abolished after intravenous injection of diazepam (5 mg). Although the screen for autoantibodies did not fit the diagnostic criteria of SPS spectrum disorders including PERM, the presence of muscle rigidity, immobility, muteness, agitation, and significant response to benzodiazepines, even in the absence of waxy flexibility and negativism, are fundamental signs for the diagnosis of catatonia. Based on their presence, a diagnosis of possible PERM and probable malignant catatonia was made.

A therapeutic regimen of oral diazepam and baclofen was initiated; diazepam at 40 mg/d restored speech, and dose escalation of diazepam to 60 mg/d nearly abolished the hyperekplexia. Since the patient continued to show agitation and difficulty with communication, we initiated a 3-day intravenous corticosteroid course (methylprednisolone, 1 g/d) on day 23 of the hospitalization, followed by oral corticosteroid therapy (prednisone, 1 mg/kg per day). Although steroid therapy slightly improved muscle rigidity, it did not have any effects on the visual hallucinations or psychomotor excitation, and had only a partial effect on muscle rigidity (mRS score, 5).

Bilateral ultrabrief pulse mECT was initiated and was administered thrice a week using a Thymatron System IV; no complications occurred. Thiamylal sodium was used as the anesthetic agent (dose range, 175–350 mg), suxamethonium chloride was used as the paralytic agent (30 mg), and flumazenil was used to reverse benzodiazepine sedation before mECT (dose range, 0.2–0.4 mg). She was treated with mECT on days 43, 45, 50, and 52 of hospitalization. After day 52, the patient's mental condition became stable. The patient's agitation, incoherence, soliloquy, stated hallucinations, and delusions diminished completely, and her ability to communicate improved dramatically. Moreover, her muscle rigidity reduced, such that she could walk around her room (mRS score, 3). Immobility, hyperreflexia, urinary incontinence, muteness, and dysphagia disappeared after 8 mECT treatments. The patient did not receive continuation or maintenance mECT. She was discharged 127 days after hospitalization, with follow-ups on an outpatient basis. At 118 days after discharge from the hospital, no complications related to ECT were observed. Cognitive function was fully maintained, and both basic and instrumental activities of daily living were preserved; however, the patient had difficulties in walking down the stairs owing to spastic gait. The patient could walk without any support to a degree such that it causes no interference with her everyday life (mRS score, 1).

Culav-Sumić et al[2] reported psychiatric symptoms accompanying SPS. Psychiatric symptoms can present themselves before the onset of typical SPS symptoms, resulting in a misdiagnosis.[2]

We believe that our diagnosis of seronegative PERM with no evidence of malignancy[3] was reasonable, given the characteristic features of rapid progressive encephalomyelitis, whole-body rigidity affecting the limbs and truncal muscles, and continuous benzodiazepine-sensitive muscle activity at rest. On the other hand, catatonia also has similar presentations, including