Palilalia due to steroid-responsive encephalopathy

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Palilalia was first described by Souques in 1908 as the repetition of syllables, words, phrases, or sentences of one's own speech.^{1,2} It is distinctive from disarticulation, reiterative aphasia, and echolalia. Critchley¹ observed that the speech tends to be uttered more and more quickly and with less distinctness, so that the latter parts may be almost inaudible—the diminuendo effect. His patients were fully aware of their palilalia, but unable to control it.¹ Critchley¹ believed palilalia was due to a disorder in the motor mechanism of speaking rather than the creation of thoughts and words themselves. Benke and Butterworth³ expanded on the idea of palilalia as a disorder of motor speech and suggested that, further, it could stem from a defect in a central prearticulatory generator area, or the control module of the articulator or motor speech that provides feedback to terminate the loop. In 1924, Sterling further divided palilalia into 2 distinct types. Type A was palilalia spasmodic or heterophonic, a “compulsive reiteration of utterance with increasing rapidity and decreasing volume,” similar to previously described cases. He added a new type B: atonic or homophonic palilalia, a “reiteration of utterances at a constant rate alternating with periods of silence.”^{4,5} We present a case of palilalia caused by a steroid-responsive encephalopathy that does not squarely fall into the previously defined categories.

Practical Implications

In encephalopathies of unclear origin, it may be useful to consider palilalia in the differential diagnosis of reiterative speech disorders.

Case report

A 55-year-old right-handed woman presented with tonic-clonic activity of the right upper and lower extremity followed by tonic-clonic activity of all extremities with loss of consciousness with several days of deficits in working memory. She had to quit her job as an executive. MRI showed lesions in bilateral caudate heads, thalami, and the right temporal lobe (figure, A and B). Diagnosed with encephalitis, she subsequently improved with steroids. One year later, she developed right hemiparesis, oral dyskinesias, diminishment in memory, and reiteration of spoken words—repeating words and phrases only, at constant volume and rate. The patient never echoed the language or actions of the examiner. All reiterations were context appropriate and not rhetorical or scatologic. The repetitions were involuntary and performed unbeknownst to the patient. The patient could be interrupted and redirected by the examiner, and could then transition into free-flowing language for a brief period of time before falling into a different reiterated phrase based on the language she had just spoken. Abstract reasoning was intact. Counting and recitation of sentences learned from memory were never reiterated. A 24-hour EEG showed mostly right temporo-parietal seizures and a few left occipital seizures. There were no ictal discharges during the uttered palilalic phrases. MRI lesions are shown in the figure, C and D. A diagnosis of Hashimoto encephalopathy was suspected based on elevated serum antimicrosomal and antiperoxidase antibodies and lack of any other etiologies (e.g., negative paraneoplastic and autoimmune panel). But given

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Figure Brain fluid-attenuated inversion recovery MRI

(A) Right temporal lesion. (B) Bilateral caudate and thalamic lesion. (C) Confluent white matter lesion involving posterior superior left frontal, left parietal, left occipital, splenium of corpus callosum extending to right parietal. (D) Involvement of left subcortex including putamen. All lesions resolved, except left medial frontal residual white matter lesion.

Supplemental Data

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prominent MRI changes and the brain biopsy results suggestive of inflammation with vasculitis, primary CNS angiitis was also considered. She was treated with 1,000 mg Solu-Medrol IV for 6 days followed by a prednisone taper, starting at 60 mg daily and tapering down by 10 mg every month. Two years later, the patient presented with status epilepticus, further diminishment in memory, and severe palilalia, with the inability to carry a conversation. When steroid therapy failed, plasmapheresis showed concomitant decrease in palilalia. Her palilalic repetition reduced from multiple times to only 2 or 3 times. She was redirectable and able to transition herself to the next part of appropriate speech, and could participate in a conversation (see audio/transcript at Neurology.org/cp). When shown a videotape of her palilalia, the patient laughed and did not show any insight into her deficits. Detailed neuropsychological testing confirmed absence of auditory working memory.

DISCUSSION

We present a case that falls along a spectrum between spasmodic heterophonic and atonic homophonic palilalia. Spasmodic heterophonic palilalic speech increases in speed, decreases in volume, and can be interrupted. Atonic homophonic palilalic speech is variable in speed, nonaccelerating, and cannot be interrupted. Our patient's mixed features weaken the dichotomous labeling of palilalia. Etiologies of palilalia are heterogeneous: postencephalitic parkinsonism, ischemia, Pick dementia, frontal lobe epilepsy, and membranous lipodystrophy. This is a case of palilalia due to a steroid-responsive encephalopathy.⁶ Palilalia as an ictal manifestation of epilepsy has been localized to the left supplementary motor area and nonictal cases to the left superior frontal gyrus, frontotemporal convexity, or basal ganglia.⁷⁻⁹ Such heterogeneous locations suggest a cortico-subcortical phonologic circuit where the left prefrontal cortex and basal ganglia are important hypernodes.³ In this case, there seems to be an open loop with the absence of auditory working memory—secondary to the left frontal lesion—driving the palilalia.

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AUTHOR CONTRIBUTIONS

Riddhi Patira: study concept and design, acquisition of data, critical revision of manuscript for intellectual content. Sarah Smith-Benjamin: critical revision of manuscript for intellectual content. V.S. Ramachandran: critical revision of manuscript for intellectual content. Eric L. Altschuler: study concept and design, critical revision of manuscript for intellectual content.

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