The Journal of Rare DISORDERS

WHISTLING SEIZURES - A UNIQUE CASE REPORT OF A RARE AUTOMATISM

Anuradha Singh, MD[1], Daniel Torres, MD[1], and Kaitlyn Lillemoe, MD[2]

[1]New York University, Langone Medical Center, New York, NY, [2]New York University School of Medicine, New York, NY

ABSTRACT

Objective: We report a unique case of a 60-year-old man with ictal whistling after being injured in a serious motorcycle accident.

Methods: Clinical presentation, neurologic examination, neuroimaging, video-EEG (electroencephalogram) analysis, and therapeutic options were defined. A review of relevant literature was also performed.

Case summary: Our patient experienced frequent complex partial seizures after the head trauma. His neurologic examination was nonfocal. Magnetic resonance imaging of the brain did not reveal any lesion. Video-EEG captured 2 stereotyped complex partial seizures with a rare ictal vocalization, *ictal whistling*. The ictal onset showed poorly sustained diffuse bitemporal theta-delta rhythms, which made it difficult to determine lateralization from the scalp EEG but favored possible right hemispheric onsets.

Conclusions: Ictal vocalizations are common in temporal lobe epilepsy, though ictal whistling has been reported in both frontal and temporal lobe cases. The very few reported cases show a strong male predominance. Clear lateralization at ictal onset was not possible in our patient, but peri-ictal and interictal discharges suggested possible right temporal lobe focus.

CASE SUMMARY

A 60-year-old right-handed Latino had a serious motorcycle accident, after which he was in a coma for 12 days. He subsequently developed posttraumatic migraines and nocturnal seizures. He reported his first seizure several years after the head trauma. His seizures were preceded by occasional auras of nausea and lightheadedness. The seizures were described as brief periods of confusion and disorientation, lip smacking, staring, pacing, whistling, strange guttural noises, and spitting. The patient reported headache and mood changes in the post-ictal period. He also had occasional tongue biting and urinary incontinence. He was experiencing 6 to 7 complex partial seizures per month, with frequent secondary generalization. His seizures would last for 5 to 10 minutes. Neurologic examination was nonfocal but revealed slow psychomotor processing and poor short-term memory.

The diagnostic workup included magnetic resonance imaging of the brain, which showed only mild microvascular disease. Inpatient video-EEG (electroencephalogram) monitoring captured 2 stereotyped complex partial seizures characterized by oral automatisms, staring, and confusion, followed by prolonged ictal whistling and complex bizarre motions of the hands and legs. Ictal onset showed poorly sustained admixed theta-delta rhythms in both temporal regions; however, there was better evolution of the ictal rhythm in 1 of the 2 seizures in the right frontotemporal region (**Figure 1**). Phenytoin was tapered off and replaced by carbamazepine XR. The patient has been seizure free on a combination therapy of levetiracetam and carbamazepine XR for almost 4 years.

DISCUSSION: Ictal whistling was first described in a case of frontal lobe epilepsy by Lazzarino and Valassi in1982.[1] Eight years later, Tan et al reported stereotyped complex partial whistling seizures in a 22-year-old man, and the seizure focus was believed to be right temporal.[2] The authors hypothesized that spread from the temporal region to the supplementary motor area accounted for the whistling automatism. A causative temporal lobe focus was further supported when Loring et al described an ambidextrous patient who had a history of febrile seizures and later developed partial epilepsy at age 3-4 years.[3] He was found to have possible left temporal horn dilatation. His case shared some common clinical semiology with our case, with prominent picking automatisms, whistling, and purposeless ambulation. A scalp EEG showed bilateral rhythmic beta, but in 2 of 3 seizures the ictal evolution was best developed in the right mesiotemporal region. Loring et al's patient underwent additional invasive monitoring with 8

subdural strips and was offered a right anterior temporal lobectomy. Not only did he not benefit from the surgery, he also developed postoperative amnestic syndrome. Therefore, temporal localization was again questioned.

In 2010, Raghavendra et al published a case series of ictal whistling during temporal lobe seizures in 2 patients.[4] The first patient was a 35-year-old right-handed man in whom video-EEG captured right temporal lobe seizure onset. In the authors' second case, a 50-year-old left-handed man, the seizures originated from the left anterior mesial temporal region. Both patients became seizure free after undergoing lateralized anterior temporal lobectomies.

The lateralization and localization of ictal whistling are poorly understood. After decades of limited localization evidence, Raghavendra et al's case series with epilepsy resolution after surgery strongly argues for the anterior temporal region. However, lateralization is still elusive, as both dominant and nondominant hemispheres were implicated in these patients. Our patient also showed possible right temporal lobe origin and was on the presurgical track, but went into remission with the initiation of carbamazepine therapy.

CONCLUSIONS

Our patient is 1 of 6 reported cases of whistling seizures in the literature. The localization of ictal whistling or musical automatisms remains unclear, as previously reported cases had both right and left hemisphere onsets from the frontal and temporal regions. We believe that our patient had a right mesial temporal focus based on the description of his auras, clinical semiology and ictal onset.

Ictal vocalizations are seen with both frontal and temporal lobe seizures. Horvath et al reported that ictal

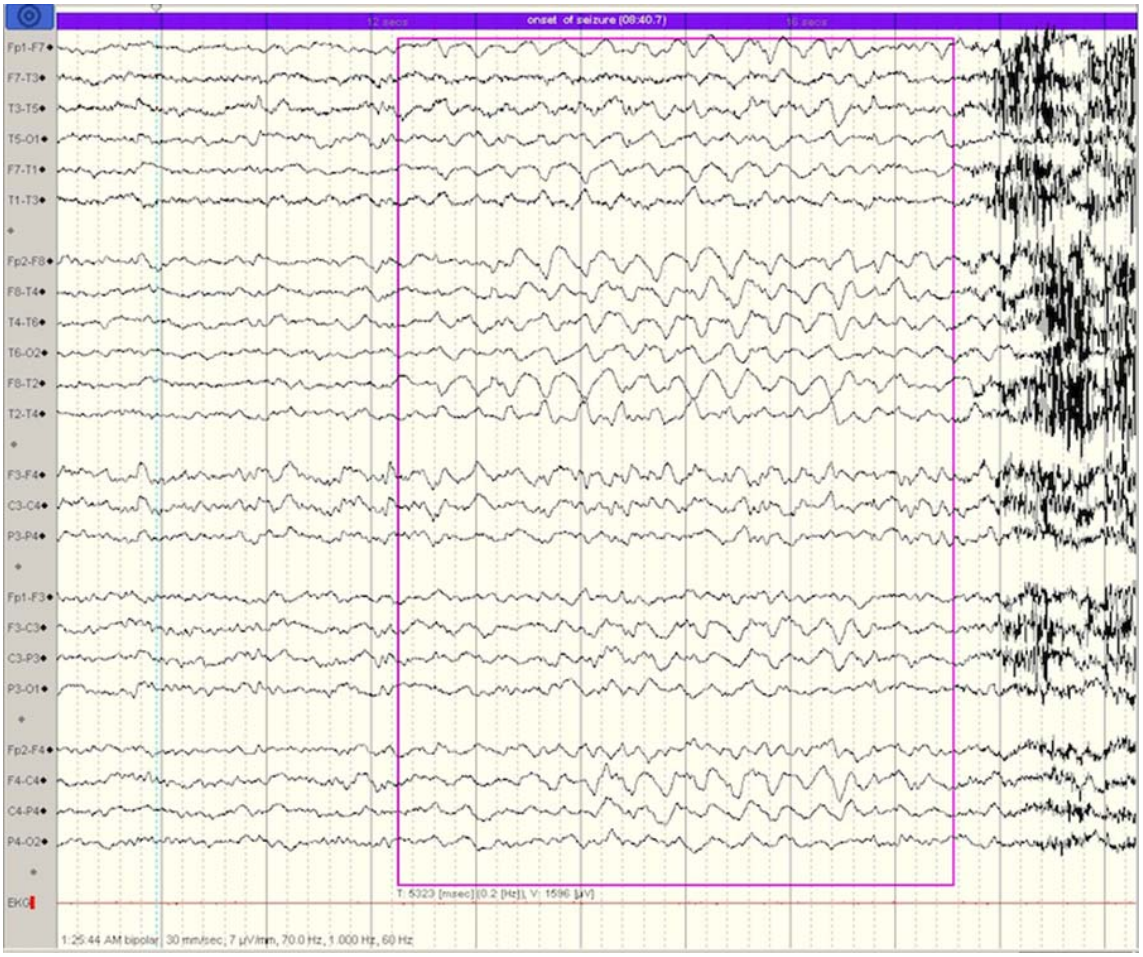

Figure 1: Ictal onset in our patient shows poorly sustained diffuse rhythmic delta rhythm in both hemispheres with slightly better field and higher amplitude in the right hemispheric leads.

vocalizations in temporal lobe epilepsy occur more often with dominant (left-sided) epileptogenic zone.[5] However, musical semiology is rare and infrequently described in the literature. Five previous cases of whistling seizures have been reported. Only 2 of the 5 cases had prolonged EEG monitoring. All cases have been males to date. There is no clear hemispheric lateralization in previously reported cases, and the ictal onsets have been either frontal or temporal . Our case was unable to show a clear lateralization, but the presence of abdominal auras, interictal data, and observed seizure semiology suggests mesiotemporal focus, possibly on the right side.

STUDY FUNDING

No industry, government, or departmental funding was granted.

DISCLOSURE

The authors have no financial relationships to disclose.

REFERENCES

1. Lazzarino LG, Valassi F. Whistling as a manifestation of epilepsy. *Riv Neurobiol.* 1982;28(1-2):127-130.

2. Tan E, Ciger A, Zileli T. Whistling epilepsy: a case report. *Clin Electroencephalogr.* 1990;21(2):110-111.

3. Loring DW, Hermann BP, Meador KJ, et al. Amnesia after unilateral temporal lobectomy: a case report. *Epilepsia.* 1994;35(4):757-763.

4. Raghavendra S, Mirsattari S, McLachlan RS. Ictal whistling: a rare automatism during temporal lobe seizures. *Epileptic Disord.* 2010;12(2):133-135.

5. Horvath RA, Fogarasi A, Schulz R, et al. Ictal vocalizations occur more often in temporal lobe epilepsy with dominant (left-sided) epileptogenic zone. *Epilepsia.* 2009;50(6):1542-1546.

Address Correspondence To:

Anuradha Singh, MD
Clinical Associate Professor,
Department of Neurology,
7W 11, 462, First Avenue,
New York, NY 10016

E-mail: anuradha.singh@nyumc.org