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Case Report

Simultaneous nonepileptic spells and nonorganic hearing loss: A case of comorbid psychogenic symptoms☆

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ABSTRACT

A twenty-eight-year-old woman with an eight-year history of partial hearing loss presented with a two-year history of worsening deafness and new-onset seizures. Evaluations of tympanic membranes, cochlea, and auditory brain stem reflexes demonstrated no physiologic basis for deafness. Video-EEG monitoring demonstrated that the patient responded normally to spontaneous auditory stimuli and that typical spells were nonepileptic in origin. Although pseudohypacusis is reported in audiology literature, and psychogenic nonepileptic spells are well-studied phenomena in neurology literature, the present case is an unusual case of dual psychogenic symptoms, a relationship that indicates poorer prognosis.

1. Introduction

Psychogenic nonepileptic spells (PNES) or pseudoseizures are paroxysmal episodes of behaviors, such as altered movement, sensation, or cognition, that, unlike epileptic seizures, occur without evidence of seizure activity determined by video-EEG [1]. Pseudohypacusis or nonorganic hearing loss (NOHL) is a condition of subjective hearing loss in the absence of any organic disease as determined by examinations, electrophysiologic tests, and neuroimaging [2,3].

We report a case of comorbid PNES and NOHL. Although PNES is well-known among neurologists and epileptologists, NOHL is usually presented to hearing specialists such as otolaryngologists and audiologists. Conversely, each phenomenon presents rarely outside of the matching subspecialty. These two psychogenic disorders have not been reported within the same patient. The case represents a juxtaposition of factitious symptoms that, in studies of patients with PNES and other comorbid, nonphysiologic complaints, implies poor acceptance of the diagnosis and poor psychological prognosis.

2. Case report

A 28-year-old woman presented with a two-year history of seizures consisting of warning symptoms of a metallic taste and scintillating scotoma in bilateral visual fields, staring, occasional left arm posturing, and, ultimately, loss of consciousness and generalized shaking. Weekly seizures (typically lasting about 1–10 min) sometimes clustered in waxing and waning episodes lasting about 3 h. Postictal symptoms included amnesia, fatigue, and diffuse headache. Urinary incontinence, tongue biting, and mild injuries or contusions occurred intermittently during or as a result of seizures. At their worst, seizures led to emergency room and inpatient admissions for which she was sometimes sedated and intubated. Treatment with phenytoin and various benzodiazepines (mainly clonazepam) was ineffective.

About six years before seizure onset, the patient reported gradual hearing impairment. Her family history was significant for a younger sibling who had congenital deafness, and the patient learned American Sign Language (ASL) at a young age. She used these skills, once professionally trained, in a career as a hearing interpreter. About two years before presentation – about the time that the seizures began – she complained of complete deafness, and she was outfitted with bilateral hearing aids.

Her psychiatric history was significant for history of physical and sexual abuse by her stepfather from the age of 8–18. She had four pregnancies as a result of sexual abuse, all resulting in miscarriages. She had been seen by a psychiatrist since the age of 16 and was diagnosed with posttraumatic stress disorder. She reported mild head trauma when she was sixteen following which she had a brief episode of loss of consciousness. She had a previous history of abdominal pain and intermittent diarrhea since the age of 13, for which she underwent inconclusive evaluation including normal abdominal CT; upper and lower GI endoscopy; and colorectal, ileal, duodenal, and stomach biopsies. She was diagnosed with possible irritable bowel syndrome versus anorexia nervosa, and GI symptoms improved over time.

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Seizures led to driving restriction and to loss of work. She was married and had two uncomplicated pregnancies at ages 19 and 21. She denied any history of exposure to toxins, loud noises, recent head or neck trauma, CNS infection, or febrile convulsions. She also denied medication use except for phenytoin; however, review of her prescription records was significant for clonazepam from multiple prescribers.

3. Results

Admission to the University of Virginia epilepsy monitoring unit revealed the patient's apparent skill at lip reading. The deaf translator reported normal use of ASL. Physical and neurological examinations were normal – including the lack of dysarthria – save for inability to hear spoken directions with or without the use of hearing aids. She had no evidence of vestibular dysfunction.

During four days of continuous video-EEG, she had multiple episodes of her typical spells – convulsions with asynchronous, migratory movements of the extremities and loss of responsiveness – with no evidence of seizure activity and preservation of waking alpha rhythm during unresponsiveness. Interictal EEG showed no interictal epileptiform discharges.

Beyond PNES, further attention to audio–video recordings during video-EEG demonstrated that the patient displayed behaviors that were unlikely to occur in the setting of complete deafness: she had brief conversations with nonmedical hospital staff that continued when the staff turned their backs and when hearing aids were not worn. She responded to indirect verbal communication. She was startled by abrupt noises occurring behind the drawn curtain of the next bed in her nonprivate room. Finally, we noted that her cell phone was set to normal ringtones and not to vibration, and pointedly, we noted that she used a cell phone in the first place.

Her evaluation for hearing loss included audiometry consultation at the University of Virginia. Despite profound hearing loss in audiograms [AD speech reception threshold (SRT) = 110 dB (normal < 25 dB, profound hearing loss >90 dB); AS SRT = not reported], she had normal tympanograms, normal otoacoustic emission, intact acoustic reflexes bilaterally on contralateral and ipsilateral testing, and intact auditory brain stem reflexes down to the level of 15 dB from both ears. Test results supported normal hearing sensitivity at least in the 1000- to 4000-Hz range. Repeat brain stem auditory-evoked potentials with square-wave stimuli at 75 dB demonstrated intact short latency potentials (Fig. 1).

She refused formal neuropsychological evaluation. Education pertaining to PNES was provided. Despite multiple sessions discussing normal test results in both realms of epilepsy and hearing both by neuropsychology consultants and by the primary team, she and her family remained unwilling to accept the diagnosis, requesting more evaluations to be performed. The patient was discharged and lost to follow-up.

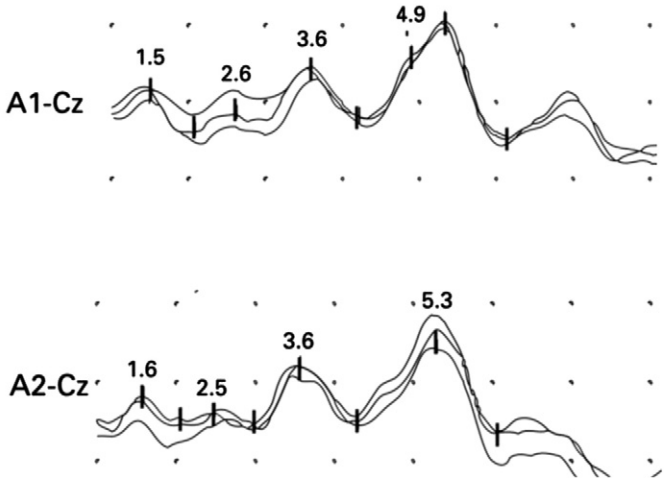

Fig. 1. Brain stem auditory-evoked potentials showed bilaterally intact short latency waveforms with normal absolute and interpeak latencies. Stimulations were performed at 75 dB and recorded from the rarefaction sweep.

4. Discussion

This case of simultaneous PNES and NOHL is important for several reasons. First, the case is unique in its combination of features, mimicking epilepsy and profound hearing loss. Second, the details aiding diagnosis – the history of sexual abuse, an eating disorder, and accumulated somatoform complaints – in combination with objective, physiologic testing were only discoverable during an extended inpatient setting; the value of inpatient video-EEG with its opportunity for in-depth history and investigation goes beyond extended monitoring, especially when patients may initially present with self-selected or incomplete histories. Third, because many findings in the evaluation of hearing loss are subjective, direct observation of patient behavior inconsistent with deafness was an important aid to the diagnosis of NOHL. Finally, patients with multiple psychogenic complaints experienced over a long period may not respond as desired to the usual recommendations in informing and treating patients with PNES.

A factor that may have contributed to delayed diagnosis was that the patient's main symptoms – early, progressive hearing loss and PNES – spanned different subspecialties. In fact, in the thirty years of accumulated experience of the UVA epilepsy monitoring unit, this patient is the only one with pseudohypacusis in our records. The simultaneous presentation of NOHL with PNES is unique, but past occurrences of NOHL (reported to occur in 13%), as well as other pseudoneurological complaints [such as blindness (18%), numbness (58%), weakness (49%), and paralysis (42%)], are reported in studies of PNES [4].

Pseudohypacusis is well-described in audiology and otolaryngology literature, a condition with a prevalence ranging between 2–9% in adults and 2–7% in children [3,5]. Nonorganic hearing loss has been reported to be more common in young women and children. Often, NOHL presents as an acute, unilateral onset. Concomitant organic hearing loss may also be present [6]. In adults, NOHL is mostly attributed to malingering [7]. In contrast to described patients, the present patient had a chronic, progressive course of hearing loss of prolonged duration. Deafness in a sibling as well as skills with ASL perhaps biased past evaluations as well as provided ample templates of deaf behavior.

Despite the unusual presence of pseudohypacusis, the present patient shared some characteristics and risk factors with other patients with PNES: female gender; age group; history of eating disorder, drug abuse or seeking; head trauma; and physical and sexual abuse [8–10]. Our patient refused formal psychiatric diagnosis. Given her GI complaints and pseudoneurological findings (but with the absence of reproductive system and pain complaints), she fell short of DSM-4R criteria for somatoform disorder but qualified for somatic symptom disorder under the DSM-5 criteria. Conversion reaction, body dysmorphia, and malingering were possibilities as well. Given that NOHL as a conversion disorder has rarely been described to persist longer than two weeks [11], factitious disorder and malingering remain as distinct possibilities [7].

Treatment of PNES begins with a forthright and clear presentation of the physiological findings backed by offers for psychological and psychiatric consultation [12–14]. In this case, the patient and her family were reluctant to accept the diagnoses. Spells persist in about one-third of patients with PNES [15,16]. Longer duration of symptoms, denial of psychosocial problems, and a diagnosis of somatoform disorder are all associated with poor outcomes [15,17–19].

In summary, somatoform abnormalities are common in patients with PNES and may indicate poor prognosis. The patient's poor acceptance of the diagnosis, refusal of psychological evaluation and treatment, and loss to follow-up all suggest that she will continue to manifest symptoms

and not enjoy improvements in quality of life, employment, and independence that patients with successfully treated PNES achieve [16].

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Conflict of interest

There is no funding or support to report. There are no conflicts of interest reported by the authors.

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