

Psychogenic Nonepileptic Seizures in a Mother and Her Son – Fate or Chance?

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Abstract

Psychogenic Nonepileptic Seizures (PNES) are defined as typical seizure-like activities or behaviors without cortical epileptiform discharges. The diagnosis of PNES remains a challenge and continuous video electroencephalography (cEEG) remains the gold standard to differentiate between epileptic seizures and PNES. Despite advances made in our understanding of PNES, diagnosing and treating this well-known entity remains a challenge. 25% of patients with seizures will be misdiagnosed as having PNES or epileptic seizures even by certified Neurologists. Do patient characteristics and demographics increase our accuracy to diagnose PNES? We present a case of a mother and her son, both diagnosed with PNES by cEEG monitoring in our epilepsy monitoring unit. While familial prevalence of conversion disorders is reported in the literature, this will be the first case report describing the incidence of PNES in two members of the same family. Do psychogenic epilepsies run in families? Will the diagnosis of PNES in one family member increase the probability of PNES in their relatives with seizures? Larger patient pools need to be studied to draw a definite conclusion regarding our observation.

Keywords: Psychogenic nonepileptic seizures; PNES; Epilepsy; Conversion disorders; Families in PNES

Introduction

Psychogenic Nonepileptic Seizures (PNES) are defined as typical seizure-like activities, behaviors or experiences without simultaneous cortical epileptiform discharges [1]. PNES include a wide spectrum of neurological symptoms, mostly perceived as seizures or fits by patients or their care-givers, without a clear underlying organic cause. It is a functional disorder which is categorized as a Conversion Disorder, or more precisely as a Dissociative Disorder under ICD-10.

Clinical cues like eye-closure for >20 sec at seizure onset, geotropism, weeping, ictal stuttering or post-ictal whispering, out-of-phase or side-to-side oscillatory movements, chaotic or disorganized thrashing, and/or absence of stertorous breathing postictally - all favor a diagnosis of PNES [2-10]. However, similarities between semiology of PNES and certain epileptic seizures, especially frontal-lobe epilepsies, need to be underscored [11-15]. Clinical findings are not very specific for any one kind of seizure disorder. Tongue biting, self-injury, and incontinence were historically perceived as signs of epileptic seizures alone. Now we know that up to 66% of patients with PNES also report similar complaints [16]. Thus, clinical signs alone have limited utility in making an accurate diagnosis of seizure subtype. Diagnosing PNES is a challenge even for an experienced epileptologist. Hence, long-term continuous video encephalogram (cEEG) monitoring is the preferred test for the diagnosis of PNES [17]. A more certain benefit is obtained when a patient experiences a typical seizure without accompanying electrographic cortical abnormalities [18].

While there is a lot written in the literature regarding the ictal features and diagnostic techniques for PNES, a little has been studied regarding the socio-demographics and familial characteristics of these patients. Hereby, we present two separate cases from a same family—both of which were diagnosed with epilepsy and treated for years before presenting to our clinic. To our understanding, this would be the first description of two patients with PNES in the same family.

Case Presentation

First, we would like to present a 37-year-old right-handed African American man who presented to our clinic with a history of seizures. He

reported his first seizure at the age of ten years and has had hundreds of such episodes since. These events were always triggered by stress. He denied any prodromal phase or aura-like symptoms. His typical episode lasted for 10 minutes and involved vigorous shaking of his head and both upper extremities. He has had falls during these events. He endorsed tongue bite during several such events, denied bladder or bowel incontinence, and denied loss of awareness of his surroundings. Post-ictal phase was marked by tiredness and confusion which lasted for minutes to hours. He was on several different AEDs in past, and most recently was treated with topiramate 25 mg BID. Routine encephalograms (EEG) were done in past but the results were not available for us to review. Medical history was remarkable for essential hypertension, morbid obesity and major depressive disorder for which he was following up with a psychiatrist, but not taking any antidepressant. Denied any major surgeries, trauma or recent hospitalizations. He had a 20 pack-year smoking history, drank beer occasionally and denied any illicit drug use. His home medications included an ACE-inhibitor and a diuretic for hypertension, and Topamax for seizures as previously noted. Family history was remarkable for seizure disorder in his mother. His neurological examination was unremarkable and non-focal. Decision was made to temporarily up-titrate topiramate to an antiepileptic dose of 200 mg twice daily with plans to admit him to epilepsy monitoring unit (EMU) for spell characterization. We had a high clinical suspicion for PNES secondary to his atypical seizure semiology. Upon holding topiramate, we initiated a continuous video electroencephalogram (cEEG) monitoring. First 2 days of recording were uneventful – clinically and electrographically. On day 3, he was subjected to sleep deprivation and seizure induction by photic stimulation and suggestibility. Then, he developed vigorous bilateral upper extremity tonic-clonic activity

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and up-rolling of eye balls which lasted for about three minutes. He remained responsive and was answering simple questions during this event. Review of cEEG recording during this event did not show any epileptiform discharges. Patient endorsed that this witnessed event was typical for his seizure-like episodes.

Incidentally, his mother also carried a diagnosis of epilepsy for 30 years and was seen in our clinic a few years back. This was a 61-year-old African American lady who was diagnosed with adult-onset seizure disorder in her thirties. Her episodes were not associated with aura or prodromal symptoms. A typical episode was described as generalized tonic-clonic activity without clear laterality which were occasionally associated with tongue biting, urinary incontinence and a post-ictal phase comprising of generalized fatigue and retrograde amnesia without recollection of the ictal events. Her episodes were usually nocturnal and hence only a few were witnessed by her children. She was treated with different AEDs in the past but was off all her AEDs at the time of presentation. At first, she would have 3-4 episodes annually. Her frequency increased and one of her episodes culminated in a motor vehicle accident. Her past medical history was remarkable for major depressive disorder and was not on active treatment. She was initially started on levetiracetam 500 mg BID which helped in reducing the frequency of her typical episodes. Brain imaging and a routine EEG were unremarkable. Levetiracetam was increased to 750 mg twice a day and cEEG monitoring in our EMU was planned to further characterize her spells. She lost her follow up with our clinic for two years because of non-medical reasons. Upon re-establishing care with us, patient was still on levetiracetam at the same dose. Her seizure semiology and frequency remained unchanged. Oxcarbazepine was temporarily added given her history of evolving depression and mood symptoms. Shortly after, patient was admitted to EMU. At admission, AEDs were held and cEEG did not show any seizure or epileptiform discharges over the first forty-eight hours. On third day, during a placebo induction using verbal suggestion, along with hyperventilation and photic stimulation, while sleep deprived, the patient had an episode that lasted greater than one minute and consisted of head shaking as well as low amplitude shaking of all extremities. The patient identified this as a typical episode. This episode was not associated with any electrographic evidence of epileptic seizure activity.

Both of our patients had been diagnosed with epilepsy for years and were treated for the same. They underwent a great ordeal of physical, psychological, financial and social stress because of this misdiagnosis of Epileptic Disorder. After a careful continuous EEG monitoring and implementation of seizure induction techniques, we were able to simulate their typical seizure-like events in an observed setting of an EMU and were able to clearly demonstrate the functional origin of their symptoms. Both of our patients were eventually diagnosed with PNES and were appropriately referred to the PNES clinic for further management. They were successfully taken off AEDs and received behavioral counselling to help them handle stressful situations better.

Discussion

The prevalence of PNES ranges from 2 to 33 cases per 100,000 persons in general population. Statistically, it is equally prevalent as multiple sclerosis or trigeminal neuralgia [19]. When considering the total pool of patients with epilepsy, around 10% of patients in outpatient setting and 20% to 40% of patients in hospital setting have PNES [1,20]. As is the case with other conversion disorders, PNES begins in young adulthood [21].

Despite the high prevalence of PNES, they are often misclassified

as epileptic seizures. Alsaadi et al., estimated that up to 25% of patients with seizure disorder are misdiagnosed even by a trained specialist [22]. This number is ought to be much higher if we put together all the diagnoses of seizure disorders made in the community. In fact, majority of the patients are treated for years on different anti-epileptic medications prior to accurate diagnosis of PNES. One review reported 72-years elapse between a patient's onset of PNES and the correct diagnosis [23]. This not only subject patients with PNES to numerous potential adverse effects of AEDs and potentially harmful interventions for pseudo-status epilepticus from repeated emergency room visits, but also lead to a great ordeal of psychological, economic and social stress. Many patients with PNES bear significant morbidity from inappropriate treatment for epileptic seizures.

PNES, when not properly diagnosed and promptly treated, is a burden for patients, families and the society. Because of a higher prevalence in younger adults and women, it disrupts the fabric of family dynamics, affects marital life and predisposes to child neglect. Wood et al., observed that families of patients with PNES reported more health problems, perceived criticism, somatization and distress than did families of patients with epilepsy [24].

The etiology of PNES is poorly understood. A genetic predisposition suggesting a neurobiological dysfunction leading to PNES has been suggested [25]. Reuber et al., put forth a multifactorial biopsychosocial model to explain the etio-pathogenesis of this disease which elucidates interplay between predisposing, precipitating, perpetuating, and triggering factors. Based on this model, an underlying genetic predisposition, if coupled with an early experience of abuse or interpersonal problems, could manifest as PNES upon encountering a stressful situation. Once manifested, it may perpetuate with repeated stressors like avoidance, misattributions, sick role, or financial disincentives [26].

Conclusion

As illustrated by our case above, when a woman in her 3rd or 4th decade develops a conversion disorder such as PNES, the improper defense mechanisms like dissociation can be transferred to her children who then progress to develop a similar disorder if they are not counselled or provided with adequate psychological support in a timely manner. This reflects a very important observation as the economic, health-care, and social burden of this transference of immature psychological defense mechanism can be enormous in modern times.

Timely and accurate diagnosis of PNES leads to a reduction of emergency visits by 97%, of clinic visits by 80%, of diagnostic testing by 76%, and medication charges by 69%, in the first 6 months, based on the observation by Martin RC et al. [27]. It also avoids a climate of increased distress, hostility, and criticism, plus a concentration on somatic aspects of functioning, which may lead to increased and more severe patterns of somatization among distressed family members as underscored by Wood et al. [24].

Is it possible that there is a strong familial component that determines the incidence of PNES? Are relatives of such patients at a higher risk of developing PNES in their lifetimes? If true, then is there a genetic explanation to such inheritance? This one report of two cases might not be sufficient evidence to draw this conclusion. However, it does incite a curiosity. Larger studies and retrospective review of familial characteristics of patients with PNES are warranted to study our observation in further detail.

Further, it would be interesting to know whether a known diagnosis

of PNES in a family member increases the pre-test probability of continuous video electroencephalography (cVEEG) in diagnosing PNES. If this association in absence of other risk factors for epileptic seizures is statistically significant, we will in fact be able to predict the diagnosis of PNES early in the disease course that will significantly reduce morbidity associated with delay in accurate diagnosis.

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