Case Report

Pseudoseizures with urinary incontinence in a child with Neuromyelitis optica: a rare association

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ABSTRACT

Background: Paroxysmal non epileptic events or pseudoseizures are common seizure mimics in children. But occasionally PNES events can present or coexist along with organic neurological disorders complicating the clinical scenario posing great challenges for diagnostic and therapeutic implications. Unusual symptomatology in PNES like frothing, tongue bite or urinary incontinence can be seen in PNES. Here, we present a rare case of pseudoseizures with urinary incontinence in a child with Neuromyelitis optica which posed a challenge for diagnosis and proper management with a favourable clinical outcome. A 14 years old girl who was known diagnosed case of Neuromyelitis optica on treatment with oral steroids presented with a history of repeated attacks of involuntary movements affecting all four limbs preceded by a feeling of fear and pain and these events were associated with urinary incontinence. Electroencephalogram and magnetic resonance imaging were performed on the patient. Ictal video EEG confirmed the diagnosis of pseudoseizures. Neuropsychological evaluation was performed which revealed multiple psychological stressors. Repeated counseling sessions and psychotherapy provided significant improvement in the form of remission of pseudoseizures.

Keywords: Epilepsy, Neuromyelitisoptica, Pseudoseizures or Psychogenic Non-Epileptic Seizures, Pseudoseizures, Urinary incontinence

INTRODUCTION

Pseudoseizures or Psychogenic Non-Epileptic Seizures (PNES) are disorders in which there are paroxysmal series of altered behavior that mimics epileptic seizures however it lacks organic causes such as dysfunction of central nervous system and the associated EEG epileptic changes. It is considered as a somatoform disorder. Pseudoseizures diagnosis can be suspected by a complete physical examination and biopsychosocial assessment. It’s incidence has been estimated to be 6.5-10.6 % in many studies and 20 % of patients visiting the epilepsy centers. But occasionally PNES events can present or coexist along with organic neurological disorders complicating the clinical scenario posing great challenges for diagnostic and therapeutic implications. Unusual symptomatology in PNES like frothing, tongue bite or urinary incontinence can be seen in PNES. Here, we present a rare case of pseudoseizures with urinary incontinence in a child with Neuromyelitis optica which posed a challenge for diagnosis and proper management with a favorable clinical outcome.

CASE REPORT

A 14 years old girl who was known diagnosed case of Neuromyelitis optica on treatment with oral steroids presented with a history of repeated attacks of
involuntary movements affecting all four limbs preceded by a feeling of fear and pain and these events were associated with urinary incontinence. The child was premorbid normal till 12 years of age after which she developed sudden onset painless loss of vision in the left eye. At that time, MRI brain and spine screen was done which showed evidence of left sided optic neuritis and longitudinally extensive intramedullary lesions in cervical cord suggestive of neuromyelitis optica. She was seropositive for anti-aquaporin 4 antibody.

During the acute phase, she was treated with high dose IV Methylprednisolone therapy. She showed improvement in visual acenty on the left eye, but she was left with optic atrophy and permanently impaired vision on the left eye. Subsequently, she was given monthly pulse of IV rituximab for 3 months. Later she switched to alternative forms of therapy. She relapsed 2 years later with bilateral painless vision loss and weakness of bilateral lower limbs. MRI brain and spine showed bilateral optic neuritis and cervical cord myelitis suggestive of NMO relapse. She was treated with high dose IV methyl prednisolone followed by oral steroids. She improved clinically. Visual acuity returned to baseline and weakness of lower limbs improved.

Nearly 2 weeks later she presented with repeated attacks of involuntary movements affecting all four limbs preceded by a feeling of fear and pain and these events were associated with urinary incontinence. These episodes mostly occurred during the night whenever she woke up. These events were preceded by a sense of intense gripping fear and pain in both lower limbs. These events were also associated with involuntary passage of urine.

MRI brain and spine was repeated which showed resolution of the lesions which raised the possibility of pseudoseizures. Oxygen saturation was 96% during the event on room air, no dilatation or tachycardia was found and plantar down going. There was spontaneous recovery of consciousness without headache, vomiting or fatigue. Ictal video EEG recorded the habitual events and then there was no ictal correlate and hence the diagnosis of pseudoseizures was confirmed.

Reason for referral to psychiatry was normal ictal EEG and presence of multiple psychosocial stressors. Episodes were triggered by suggestibility. There was pervasive anxiety and apprehension about recurring episodes. Neuropsychological evaluation was performed which revealed parental marital disharmony, learning issues, mental stress and fear due to her medical condition and sibling rivalry, projective test was done using thematic perception test. Mother and child in hostile and fearful environment.

Individual therapy with the child focused on establishing rapport, distraction and deep breathing technique. Family sessions focused on psychoeducation about psychological model for seizure like phenomenon, reducing secondary gain in the form of attention and over involvement during episodes. Sibling rivalry was addressed through joint session with sibling and mother with focus on conditional positive regard and differential reinforcement. Child seems to have writing, spelling and arithmetic difficulties and advised to take individualized learning methods. Patient was started on tab clonazepam 0.25 mg three times a day and it was given in view of persisting anxiety. She was continued on oral azathioprine for maintaining remission in neuromyelitis optica. Significant improvement was noticed in the form of reduction of frequency from five episodes during day and ten episodes during night to three in a day and two in the night.

**DISCUSSION**

The description of paroxysmal alterations in behavior that looks like epileptic seizures is called pseudoseizures but lacks any organic cause. Pseudoseizures or Psychogenic Non-Epileptic Seizures (PNES) accounts for 17% to 30% of patients admitted to epilepsy units. The patient has normal laboratory tests and physical examination. Diagnosis of pseudo seizures must be precisely recognized because mistake in diagnosis can be harmful. Actually it is very difficult to diagnose pseudoseizures due to presence of overlapping in syndromes of true epilepsy with pseudo epilepsy. There are many characteristics of pseudo seizures including absence of tongue bite, absence of urinary incontinence, normal pupillary reflex and normal pupil size, whereas it is dilated in organic seizures. Other features of pseudo seizure include occurrence of seizure like phenomenon at all times, increased frequency in the presence of family members or people associated with conflict and flexor plantar response. Our patient fulfilled above criteria for pseudo seizure presentation. she did not have tongue bite and most of the episodes happened in front of treating team and family members. She was suffering from repeated attacks of involuntary movements affecting all her four limbs, but during the event she was aware of her surroundings and she regained full consciousness and function after the event.

In a case series of 63 adults patients by Patidar et al, one patient had a neurological morbidity in the form of Neuromyelitis optica. In a study by Nakano et al, it was concluded that patients with demyelinating illnesses like MS/NMO are at higher risk of epileptic seizures when compared to normal individuals. To the best of our knowledge, this case is the first report of pseudoseizures complicating a child with neuromyelitis optica.

Presence of urinary incontinence and few episodes happening during sleep were perplexing in this case. Bladder incontinence and tongue biting are not exclusive to epileptic seizures, since they occur separately or together in about one half of EEG-proven pseudoseizures. PNES may be stereotyped, resulting in significant injury, there may be tongue biting, urinary incontinence, colour...
and respiratory changes. Episodes occurring from sleep are reported (apparent sleep when actually recorded with EEG). Recording the habitual events by Video EEG and demonstrating the non-epileptic origin of seizure is the gold standard method of diagnosis.

Electroencephalogram (EEG) may be normal in 30% of epilepsy patients, however it is important to be performed during the seizures to establish a correct diagnosis. In the present case, normal EEG was recorded which demonstrated a normal background with no abnormal focal slowing or epileptiform discharges.

In the current case scenario, the stressors identified for possible overlay with pseudo seizure include sibling rivalry, Interpersonal conflict between parents and achievement-capacity mismatch. The child had significant improvement in the symptomatology, which points toward better prognosis. Considering the complexity of family dynamics and gain out of the symptom to deal with conflict related anxiety, chances of recurrence is likely. Reinforcement of her symptoms by secondary gain from family is also likely to lead to relapse. In child there are certain clinical features which are specific to pseudo seizure in children and adolescent. These include poor socio economic status, low IQ, presence of psychiatric comorbidity, predominantly motor symptoms, delayed diagnosis and guarded prognosis.

So the above case describes a prototype presentation of seizure-pseudo seizure overlay in a case of neuromyelitisoptica. It is imperative to elicit clinical history and sequence of events with careful exploration of psychological conflicts whenever there is suspicion of pseudoseizures overlay.

CONCLUSION

Patients with PNES are commonly misdiagnosed as having epilepsy. There may be considerable delay before correct diagnosis and as a consequence, patients are exposed to inappropriate AED treatment and are at risk of iatrogenic harm from inappropriate interventions for uncontrolled 'seizures'. Diagnosis is best confirmed by VEEG. This case highlights coexistence of serious neurological disorders like neuromyelitisoptica with pseudoseizures and also atypical features like urinary incontinence and nocturnal events (occurring out of pseudo sleep) does not exclude the diagnosis of pseudoseizures.

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