Case Report

Recurrent prolonged fugue states as the sole manifestation of epileptic seizures

Geeta A. Khwaja, Ashish Duggal, Amit Kulkarni, Neera Chaudhry, Meena Gupta, Debashish Chowdhury, Vikram Bohra

Department of Neurology, Govind Ballabh Pant Hospital, Delhi, India

Abstract

A fugue state is defined as an altered state of consciousness with varying degrees of motor activity and amnesia for the event. It may last for hours to days and may be psychogenic or organic in nature. Epileptic fugue states can be encountered in patients with absence or complex partial nonconvulsive status epilepticus or may occur as a postictal phenomenon in patients with generalized seizures. “absence status epilepticus” (AS) is rare and seen in only 2.6% of the cases with “childhood absence epilepsy” (CAE). The diagnosis of AS can be elusive, but sudden onset and termination of the fugue state, classical electroencephalogram (EEG) features, and response to a therapeutic trial of benzodiazepines helps in confirming the diagnosis and differentiating it from nonepileptic fugue states. We report a childhood onset case, with a 10 years history of recurrent episodes of prolonged fugue state lasting for up to 24 h, as the sole manifestation of epileptic seizures. The EEG features were suggestive of an AS, but there was no history of typical absences, myoclonus, or generalized tonic clonic seizures. This unusual and rare case cannot be categorized into one of the defined epilepsy syndromes like CAE but belongs to a recently identified syndrome of idiopathic generalized epilepsy known as “Absence status epilepsy” in which AS is the sole or the predominant seizure type.

Key Words

Absence status epilepticus, fugue state, idiopathic generalized epilepsy

Introduction

Fugue state, also referred to as twilight state, is an altered state of consciousness with varying degrees of motor activity and amnesia for the event. It may last for hours or days and can be encountered in various neurological and psychiatric disorders. Epileptic fugue states can be encountered in patients with absence or complex partial nonconvulsive status epilepticus or may occur as a postictal phenomenon in patients with generalized seizures. We report an unusual case with childhood onset “recurrent prolonged fugue states” due to “absence status epilepticus” (AS) as the sole manifestation of epileptic seizures.

Case Report

A 15-year-old female, presented with 10 years history of recurrent episodes of acute onset, prolonged confusional states, accompanied by excessive somnolence, lasting for 15-24 h or more. The patient was a full-term normal delivery, with normal developmental milestones. The attacks first manifested at 5 years of age with a frequency of 1-2 per month, the current frequency being 2-3 per month. The attacks were stereotyped in nature and characterized by a prolonged fugue like state which would descend all of a sudden without any prior warning. During the attacks, the patient would appear dazed and become clumsy and ataxic. She displayed excessive sleepiness and remained confined to bed most of the day but remained partially responsive, answering in monosyllables, drinking and eating food when offered and would also attended to her toilet needs with assistance. The attacks were not accompanied by any jerks, tonic-clonic contractions, automatisms, or incontinence. The attacks would terminate as suddenly as they started and the patient would then resume her normal day to day activities as before. Interictically, her behavior was normal but her overall school performance was poor. There was no history of absence attacks, complex partial seizures (CPS), myoclonic jerks, or generalized tonic-clonic seizures. There was no
history of head injury or family history of seizures. She had received valproate (600 mg/day) for 7 years following the onset of these attacks without any significant change in the frequency or duration of the attacks.

On examination, at the time of presentation, she was fully conscious and alert. Her minimum mean square error score was 27/30. On detailed higher mental function assessment, her fund of knowledge was poor and frontal lobe performance (literal fluency, mental flexibility, motor programming) was impaired. New learning ability, verbal memory tasks, and constructional ability was also impaired. There was no motor or sensory deficit.

Routine investigations including haemogram, blood biochemistry, and magnetic resonance imaging brain were normal. An interictal electroencephalogram (EEG) done at 8 years of age, showed normal background activity with frequent intermittent short lasting bursts of generalized 2.5-3 Hz spike wave discharges lasting for 4-s [Figure 1]. An ictal EEG done at 12 years of age revealed continuous generalized 2.5-3 Hz spike wave discharges throughout the trace without any discernable background activity [Figure 2]. A current EEG done 2 days following the last attack, revealed 3-4 Hz slow background activity with frequent intermittent bursts of generalized, bilateral symmetric and synchronous, high voltage slow wave activity intermixed with spike discharges and at places 3 Hz spike and wave discharges suggestive of a generalized seizure disorder [Figure 3]. Video EEG monitoring in the immediate postfugue state did not reveal any evidence of typical or subtle clinical absence attacks.

On the basis of the history and EEG findings, a diagnosis of prolonged fugue states or AS as the sole manifestation of epileptic seizures was made. In view of a history of lack of response to valproate, she was started on levetiracetam (1.5 g/day). Lacosamide (100 mg/day) was added subsequently because of persistence of epileptiform discharges in EEG after 1 week. There was, however, no decrease in frequency of attacks even after 1 month. However, on adding clobazam (30 mg/day) during one of the fugue states she became more responsive with improvement in her sensorium and the duration of attack was significantly reduced to 6 h. Subsequently, clobazam was continued and she remained seizure free over a 4 months follow-up period.

**Discussion**

Fugue or confusional states (ambulatory states of altered consciousness) can be organic or psychogenic in nature and...
can be encountered in patients with epilepsy, dementia, head trauma, brain tumors, and various psychiatric disorders including depression, schizophrenia, mania, drugs, alcohol, and substance intoxication or withdrawal.[1] “Psychogenic” or “dissociative” fugue state may be the result of some traumatic loss, bereavement, or psychiatric disease. There may be a partial to total confusion about identity or assumption of a new identity with amnesia for the past. Prolonged fugue states lasting for days to months or states in which complex “intelligent” activities are accomplished are more likely to be nonepileptic in origin.[3] An “epileptic” fugue state can be defined as a pathological state of altered consciousness in which motor behavior may be purposeful but automatic. It usually appears during an absence or complex partial nonconvulsive status epilepticus or may occur as a postictal phenomenon in patients with generalized seizures.[3] The clinical and EEG profile in our case was suggestive of recurrent attacks of AS. During these prolonged epileptic fugue states, our patient, although conscious, appeared confused, minimally responsive, clumsy, and lethargic.

Charcot in 1889,[4] described a case with multiple, prolonged, ambulatory fugue states presumed to be epileptic in origin. Gastaut et al., in 1956[5] reported a case of a 56-year-old nurse, who had a prolonged amnestic fugue states during which she lived outdoors in the hills. The possibility of an acute psychosis was ruled out by EEG, which showed left temporal ictal discharges. Similarly a fugue like state with clouding of consciousness of varying intensity may be seen during AS and may persist for hours to days.[6][7] It may consist of a subtle, simple slowing of thought processes and expression recognized by the patient only. A frank confusional state with disturbance of alertness, attention, memory, judgment, language, anosia, and apraxia is, however, more common. Language is reduced to fragmented, hesitant, and at times irrelevant responses interrupted by long pauses associated with echolalia and palilalia. Patients are usually unable to follow complex commands. They may, however, follow simple commands, often after repeated requests, with a slow and delayed response, as was also evident in our case. The spontaneous duration of episodes of AS is variable and may range from about half an hour to several weeks. Most episodes last from 6 to 72 h, only exceptionally exceeding 1 week.[8] In typical cases, cessation of AS is spontaneous and sudden, with a striking clinical improvement, as was evident in our case also.

Clinically, four types of AS may be recognized: Typical AS, atypical AS, AS with focal features, and de novo AS of late onset.[7] “Typical AS” occurs as part of idiopathic generalized epilepsy and has an excellent prognosis. It is characterized by a simple confusional state with rhythmic 3 Hz spike-and-wave discharges on EEG. “Atypical AS” on the contrary occurs in patients with symptomatic or cryptogenic epilepsies. It is characterized by a fluctuating confusional state with more prominent tonic and/or myoclonic and/or lateralized ictal manifestations as compared to typical AS. Most of the patients have associated moderate or severe learning defect and an abnormal background on interictal EEG. The immediate prognosis is guarded with a high recurrence rate and resistance to medications. “AS with focal features” occur in subjects with preexisting or newly developing localization-related epilepsy; most often extratemporal EEG shows bilateral but often asymmetric ictal discharges. The prognosis is reportedly poor particularly in critically ill elderly patients. “De novo AS of late onset” is seen in middle-aged or elderly subjects with no previous history of epilepsy. Toxic or metabolic factors may lead to precipitation of seizures particularly in patients with psychiatric illness or those on multiple psychotropic drugs. Our patient, however, had the clinical and EEG features of de novo and recurrent AS starting in early childhood.

The diagnosis of AS is difficult and the condition is often misdiagnosed.[9] The altered mental status of AS may be wrongly attributed to a metabolic disturbance or to excessive psychotropic drug use or withdrawal. An important historical clue to the epileptic nature of confusional or fugue state is sudden onset and termination of the fugue state as was seen in our case also. EEG confirms the ictal nature of confusion and settles issues of differential diagnosis such as psychogenic dissociative fugue state and substance abuse. The essential EEG feature of AS is a bilateral, synchronous, symmetric paroxysmal activity that is unreactive to sensory stimulation. Characteristically, EEG tracings show continuous trains or frequently repeated bursts of polyspike-and-slow-wave complexes or slow spike-and-wave complexes that are diffuse, rhythmic, and nonreactive. The spike and wave activity occurs at a rate of 3 Hz or 1 to 2.5 Hz.[8] Occasionally, the spike-and-wave activity may be unusually rapid, from 4 to 6 Hz or unusually slower than 1 Hz.[11][12] In our case, the ictal EEG revealed generalized 2.5-3 Hz spike and wave activity suggestive of AS, while the interictal EEG showed normal background activity with intermittent 3 Hz spike and wave discharges. Although no clear correlation exists between the degree of altered consciousness and the EEG, alterations in consciousness are more commonly associated with the pattern of continuous, rhythmic, 3 Hz spike and wave activity as was seen in our case also.[13] A therapeutic trial of intravenous benzodiazepine if successful results in normalization of the EEG and disappearance of the confusion and is mandatory to confirm the ictal nature of the episode. Termination of the attack may be dramatic or immediate but may take minutes to hours in elderly patients with fugue state.

Although clinical manifestations in our case started at the age of 5 years, there was no history of CPS, typical absence seizures, myoclonic jerks, or generalized tonic-clonic seizures and phantom absences were ruled out by video EEG monitoring. On the basis of semiology alone, this case cannot be classified into any of the defined seizure types or epilepsy syndromes defined in the international classification of epileptic seizures and epilepsy, but the generalized 2.5-Hz spike and wave EEG discharges seen during the interictal as well as ictal stage were suggestive of an AS. As only prolonged fugue states were the sole manifestation of epileptic seizures in our case, it is unlikely to represent a case of childhood absence epilepsy (CAE). Moreover, AS is rare in patients with CAE and is seen only in 2.6% of the cases and some consider AS as an exclusion criterion for childhood epilepsy.[14] Without a history of typical absence seizures, de novo or recurrent prolonged attacks (15-24 h) of AS as the sole manifestation of idiopathic generalized epilepsy, is extremely rare but was documented in our case. Our patient most likely represents a case of “absence status epilepsy” (ASE), a new entity described by some authors in the recent past.[15]
Genton et al.,[13] in 2008 coined the term “ASE” for patients with recurrent, unprovoked typical AS, as the main clinical feature. Their patients had no family history of epilepsy and neurological evaluation and neuroimaging were normal, as was observed in our case also. Interictal EEG showed generalized spike and polyspike-wave discharges on a normal background. The response of AS to intravenous benzodiazepines was variable, but good seizure control was achieved with valproate and other anti-absence drugs. Although our patient did not respond to valproate, levetiracetam and lacosamide; she responded well to clobazam, much like one of the patients described by Genton et al. Patients with recurrent AS, as a unique, predominant seizure type have also been described by Andermann, Baykan et al., Zambrelli et al., and Bauer et al.[16-19]

The present case highlights the fact that prolonged epileptic confusional or fugue states may be the sole or predominant seizure type in patients with epilepsy. Sudden onset and termination of the fugue state offer a clinical clue to diagnosis of AS which can be confirmed by classical EEG features and a therapeutic response to benzodiazepines. In conclusion, it is important to be aware of the newly recognized syndrome of “ASE” in which recurrent prolonged attacks of AS are the sole or predominant seizure type.

References
1. Akhtar S, Brenner I. Differential diagnosis of fugue-like states. J Clin Psychiatry 1979;40:381-5.
2. Loewenstein RJ. Psychogenic amnesia and psychogenic fugue: A comprehensive review. In: Tasman A, Goldinger SM, editors. Review of Psychiatry, Vol. 10. Washington, DC: American Psychiatric Press; 1991. p. 189-222.
3. Tatum WO, Kaplan PW, Jallon P. Epilepsy A to Z A Concise Encyclopedia, 2nd ed. New York: Demos Medical Publishing; 2009. p. 141.
4. Goetz C. Chart at the Salpetriere: ambulatory automatisms. Neurology. 1987;37:1084-8.
5. Gastaut H, Roger J, Roger A. The significance of certain epileptic fugues; concerning a clinical and electrical observation of temporal status epilepticus Rev Neurol 1956:94:298-301.
6. Gastaut H. Dictionary of epilepsy. Geneva: World Health Organization; 1973. p. 63-4.
7. Thomas P, Zifkin B, Andermann F. Absence status. In: Wasterlain CG, Treiman DM, editors. Status Epilepticus, Mechanisms and Management, 1st ed. London, England: The MIT Press Cambridge; 2006. p. 91-108.
8. D’Agostino MD, Andermann F, Dubéau F, Fedi M, Bastos A. Exceptionally long absence status: Multifactorial etiology, drug interactions and complications. Epileptic Disord 1999;1:229-32.
9. Rohr-Le Floch J, Gauthier G, Beaumanoir A. Confusional states of epileptic origin: value of emergency EEG. Rev Neurol 1988;144:425-36.
10. Goldmann JW, Glastein G, Adams AH. Adult onset absence status: A report of six cases. Clin Electroencephalogr 1981;12:199-204.
11. Fujisawa T, Watanabe M, Nakamura H. A comparative study of absence status epilepticus between children and adults. Jpn J Psychiatry Neurol 1988;42:497-508.
12. Hersch EL, Billings RF. Acute confusional state with status petit mal as a withdrawal syndrome and five year follow-up. Can J Psychiatry 1988;33:157-9.
13. Shorvon S. Absence status. In: Status Epilepticus: Its Clinical Features and Treatment in Children and Adults. Cambridge, United Kingdom: Cambridge University Press; 1994. p. 76-84.
14. Panayiotopoulos CP. Absence epilepsies. In: Engel JJ, Pedley TA, editors. Epilepsy: A Comprehensive Textbook. Philadelphia: Lippincott-Raven Publishers; 1997. p. 2327-46.
15. Genton P, Ferlazzo E, Thomas P. Absence status epilepsy: Delineation of a distinct idiopathic generalized epilepsy syndrome. Epilepsia 2008;49:642-9.
16. Andermann F, Robb JP. Absence status: A reappraisal following review of thirty-eight patients. Epilepsia 1972;13:177-87.
17. Baykan B, Gokyigit A, Gurses C, Eraksoy M. Recurrent absence status epilepticus: Clinical and EEG characteristics. Seizure 2002;11:310-9.
18. Zambrelli E, Terzagli M, Sinforiani E, Manni R. Non-convulsive status epilepticus and generalised tonic-clonic seizures persisting in old age in a patient with idiopathic generalised epilepsy: A long-term observation. Neurol Sci 2006;27:436-8.
19. Bauer G, Bauer R, Dodesberger J, Benke T, Walser G, Trinka E. Absence status in the elderly as a late complication of idiopathic generalised epilepsies. Epileptic Disord 2007;9:39-42.

How to cite this article: Khwaja GA, Duggal A, Kulkarni A, Chaudhry N, Gupta M, Chowdhury D, et al. Recurrent prolonged fugue states as the sole manifestation of epileptic seizures. Ann Indian Acad Neurol 2013;16:561-4. Received: 07-03-13, Revised: 21-04-13, Accepted: 14-06-13 Source of Support: Nil, Conflict of Interest: Nil

Announcement

iPhone App

A free application to browse and search the journal’s content is now available for iPhone/iPad. The application provides “Table of Contents” of the latest issues, which are stored on the device for future offline browsing. Internet connection is required to access the back issues and search facility. The application is Compatible with iPhone, iPod touch, and iPad and Requires iOS 3.1 or later. The application can be downloaded from http://itunes.apple.com/us/app/medknow-journals/id458064375?ls=1&mt=8. For suggestions and comments do write back to us.