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Recurrent absence status epilepticus: clinical and EEG characteristics

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In order to outline the clinical and EEG characteristics of recurrent absence status epilepticus (ASE), eight cases with more than two attacks of ASE were studied. Their current ages were between 13 and 84 years, and five of the patients were women. There was a history of epilepsy in five of the patients before the first ASE episode. A varying degree of confusion was the main clinical symptom with associated mild motor signs like perioral, eyelid and generalised myoclonus, seen in one, two and four patients respectively. Two of the patients had juvenile myoclonic epilepsy. One patient had an atypical form of childhood absence epilepsy characterised by recurrent ASE attacks on awakening. There were two patients with phantom absences and late onset generalised convulsions, one patient with perioral myoclonia and absences, and finally two patients with eyelid myoclonia with absences, which are proposed syndromes. On the EEGs that revealed the diagnosis of ASE, there was a marked variability of the generalised multispike and wave discharges. The EEG findings appeared to be syndrome-related with some exceptions. IV Clonazepam lead to a dramatic improvement. Our study shows that the majority of recurrent ASE cases do not fit into the International syndrome classification.

Key words: nonconvulsive status epilepticus; absence status epilepticus; EEG; epilepsy; nonconvulsive seizure.

INTRODUCTION

Nonconvulsive status epilepticus (NCSE) is characterised by a slowness of behaviour and confusion accompanied by continuous electroencephalographic seizure activity[1,2]. Yet, it is not known whether NCSE causes any brain damage[3–6]. Absence status epilepticus (ASE), the generalised form of NCSE is usually known as an epileptic state with good prognosis, but there are only a few studies about its recurrence and syndrome diagnosis[7–10]. EEG is an indispensable diagnostic tool for the diagnosis of ASE in dealing with variations of abnormalities.

Although cases with ASE as a single recurrent seizure type have occasionally been reported, there are no systematic studies about the recurrent ASE cases[8]. One recent study has attracted attention on this issue[7]. Although it has been reported that ASE may be seen in idiopathic generalised epilepsies (IGE), there is limited knowledge about which epileptic syndromes may be closely related with this state[7,8,10].

In this study, our aim is to outline the clinical and EEG characteristics of epileptic patients with more than two clinically and electroencephalographically proven ASE attacks, and to classify these cases according to the known and 'proposed' epileptic syndromes.

PATIENTS AND METHODS

Within a period of 15 years, 29 NCSE cases were admitted to our department. The patients with lateralising clinical or laboratory signs were excluded from this study. There were 16 ASE cases showing alteration in cognition and behaviour associated with clearly symmetric, bilateral epileptic activity on EEG during this state, and improvement with antiepileptic drug therapy. At the time of the EEG showing ASE, blood biochemistry parameters were normal in all of the patients and there was no history of intoxication or drug withdrawal in any of the cases. Out of these 16 patients, eight patients who had more than two clear-cut clinically and

electroencephalographically proven ASE attacks were included in this study. All of these eight patients had a follow-up of at least two years. The clinical and laboratory features of these eight cases were reviewed in detail.

The EEGs were read and interpreted by two experienced electroencephalographers (AG, BB) in joint sessions. EEG recordings were performed with scalp electrodes placed according to the International 10–20 system with both bipolar and referential montages. Standard activating procedures were performed in all patients. The ictal as well as interictal EEGs and video-EEGs of these patients were evaluated regarding the morphology, continuity, regularity and frequency and associated clinical findings. The background activity, effects of hyperventilation, intermittent photic stimulation procedures and the effects of eye opening and eye closure were all noted as well as the responsiveness to IV Clonazepam, during the ASE episode. All but one had neuro-imaging studies. Ring chromosome 20 investigations disclosed normal results in four patients. Detailed neuropsychological evaluation could be done in four of them.

The patients were classified according to the epileptic syndromes suggested by ILAE in 1989[11].

RESULTS

The clinical findings are outlined in detail in Table 1. The current ages were between 13 and 84 years and five of the eight patients were women. The mean age at the first ASE episode was 16.5 ± 9.68 years. Five patients were known as epileptics before the diagnosis of ASE, while three had their first epileptic attack as an ASE. The mean age at the first seizure was 11.69 ± 7.27 years.

A varying degree of confusion was the main clinical symptom with associated mild motor signs like perioral, eyelid and generalised myoclonus seen in one, two and four cases, respectively.

The descriptions of the ASE episodes by the patients and their relatives were as follows:

Case 1: Disoriented in place, walks, cannot go home, confused, purposeless movements, prominent blinking, sometimes head movements, can speak, visual illusion (she says that she sees the curtains as very small) complains of headache (misdiagnosis: migraine equivalent!!), does not eat anything.

Case 2: Disoriented in place and time, walks in a dazed condition, cannot go to her classroom, could not communicate properly, cannot remember, random jerking, looks upwards.

Case 3: Dizziness, repetitive jerking, sometimes cannot eat and dress himself, interruptions in speaking, cannot go to work, describes the unusual feeling like electricity in the brain, sleepiness.

Case 4: Slurred speech, blinking, confusion, sleepiness, slowness in movements, cannot write.

Case 5: Describes the episodes as 'ending of her intelligence', forgetfulness, dullness, sleepiness, elevated appetite, increased movements, awareness of her attack, (came to hospital on her own).

Case 6: Confusion, marked perioral rhythmic movements, forgetfulness, urinary incontinence, (her mother says: 'he would not understand even if you hurt him'), swallowing difficult because of perioral movements.

Case 7: Hears but unable to answer questions appropriately, slowness of judgement and planned movements, feels like a 'sleepwalker', continuous eye and lid movements in upward direction, unable to close or open her eyes.

Case 8: Awakening difficult lasting 3–5 h 1–2 times weekly, urinary and faecal incontinence during these episodes, slowness in behaviour, blinking, myoclonia in the face and arms.

All patients were neurologically normal except two who had borderline intelligence and neuro-imaging studies disclosed normal results except mild ventricular enlargement in case 6 and a colloidal cyst in case 1 which were interpreted as coincidental findings. Ictal single photon emission computed tomography (SPECT) and hexa-methyl-propylene-amine-oxim (HMPAO) of case 8 demonstrated generalised hyperperfusion when compared with the interictal SPECT of the same patient.

ASE recurred 3–6 times in three patients, whereas in the remaining five patients the recurrences were more frequent (more than 20 to 100). The duration of ASE varied from 20 min to 5 days as seen in Table 1. Non-specific infections were found to be the most common precipitating events, mentioned in six of the patients. A temporal relationship with generalised convulsions was demonstrated in five patients in some of the ASE attacks. We observed prominent diagnostic delays and misdiagnoses in six of the patients. In one of these patients (case 3), the ASE could be diagnosed with the video-EEG examination of the patient during this state. Although he had complained of these episodes, we had interpreted those as a depressive disorder and as side effects of the treatment. EEG and video-EEG findings are shown in Table 2.

EEG findings during ASE attacks

On the EEGs that revealed the diagnosis of ASE, there were generalised multispike-wave discharges in all of

Table 1: Clinical features of the recurrent ASE cases.

No. Sex	1 F	2 F	3 M	4 F	5 F	6 M	7 F	8 M
Current age	20	25	27	28	84	17	34	13
Age at first seizure	7	17	17	18	20	1.5	10	3
Other seizure types (age at onset in parentheses)	*abs (8)	myo (23); GTCS (17)	myo (17) GTCS (17) following myo.	Rare GTCS (18) ph. abs (?)	Rare GTCS, (20) ph. abs (?)	#abs (1.5) GTCS (16)	*abs (10)	abs (6; after treatment of ASE)
Follow-up (years)	12	2	9	9.5	12	2	6	7
Neurological examination	N	N	N	(Borderline IQ) N	N	(Borderline IQ) N	N	N
Neuroimaging (CT/MRI)	MRI: colloid cyst	CT:N	MRI:N	MRI:N	Nd	Moderate ventricular enlargement	CT: N	MRI: N
History	N	N	N	Febrile convulsion, Bell's palsy	Hypertension, left heart failure	N	N	Difficult delivery
Family history	N	N	Epilepsy in aunt; Neuro-Behçet disease in the brother	N	N	N	Childhood absence epilepsy in her nephew	Epilepsy in the mother
Age at first ASE	8	17	23	18	29	7 (?)	27	3
Number of ASE attacks	6 (none after treatment)	3	>20	>20	>50	>100 (1–2/week)	3	>100 (1–2/week)
Duration of ASE (min–max)	20 min–4 h	1–4.5 h	3 h–3 days	30 min–5 days	1–2 days	30 min–6.5 h.	6–24 h	3–5 h
Precipitating factors of ASE	Infection	Infection	Sleep deprivation	Menses, drug discontinuation, infection	None	Infection	Infection, allergy treatment	None
Temporal relation with GTCS	No GTCS	+ (terminate in GTCS)	+	+ (terminate in GTCS)	+/−	+	No GTCS	No GTCS
Treatment	CLZ 3 mg + ETS 500 mg Now 0.5 mg CLZ only	VPA 1000 mg	3000 mg VPA, 180 mg PB, 4 mg CLZ, LTG 50 mg	750 mg VPA	3 mg CLZ +500 mg VPA (She did not use regularly)	400 mg CBZ worsened; 1000 mg VPA now	500 mg ETS; then 500 mg VPA	ETS 500–750 mg Now: no treatment
Outcome	Seizure-free for past 2.5 years	No ASE or GTCS with treatment; rare myo.	Poor outcome, frequent seizures	With AED treatment no ASE, ph. abs only	Without treatment frequent ASE	With AED treatment no ASE, mild abs.	With AED treatment no ASE mild abs	Seizure free for past 3 years
Misdiagnosis or diagnostic delay	Diagnosed as migraine equivalent!	Diagnosis delayed 5 years	Diagnosis delayed 1 year	None	Diagnosis delayed 40 years	Diagnosed as partial epilepsy	Not recognized in the emergency unit	His family did not recognize the ASE

Table 1: Continued.

No. Sex	1 F	2 F	3 M	4 F	5 F	6 M	7 F	8 M
Occupation	Student (not very successful in the school)	Student, married	Civil servant	Worker	Retired teacher, had 3 children	Worker (He could not attain to high school)	Housewife	Student
Psychiatric problems	None	None	Problems in work, depression	Depression and personality disorder	None	None	None	Personality disorder (problems in the school)
Ring 20	Nd	(−)	(−)	(−)	Nd	Nd	Nd	(−)
Neuro-psychological testing (in the interictal period)	*Very mild* attention deficit, frontal signs and visuo-spatial deficit	Nd	Mild attention deficit, moderate frontal signs	Moderate attention deficit, mild memory defects in recall	Nd	Nd.	Nd	Mild attention deficit, mild frontal signs and visuo-spatial deficit

*abs: Absences with eyelid myoclonus; #abs: Absences with perioral myoclonus; ph. abs: Phantom absence seizures; Nd: not done; N: normal; myo: myoclonus; ASE: absence status epilepticus; AED: antiepileptic drugs; GTCS: generalised tonic–clonic seizures. (CLZ: clonazepam; ETS: ethosuximide; VPA: valproate; PB: phenobarbitone; LTG: lamotrigine; CBZ: carbamazepine.)

the patients (Figs 1 and 2). These discharges were continuous in three patients, whereas in the others there were normal intervals lasting 1–6 s. In five patients epileptic abnormalities were irregular, in the others almost rhythmic delta waves and polyspikes were seen. The frequency of the spike and wave discharges ranged from 1.5 to 3.5 Hz, in two patients it slowed down to 1.5 Hz in some parts of the recording. The number of spikes per slow wave was greater than one in all of the EEGs with recurrent ASE. In all of the ASE cases IV Clonazepam induced a dramatic improvement of clinical and/or EEG findings (Fig. 1).

Video-EEG during ASE disclosed a confusional state and difficulty in responding and counting in three patients. In one of the patients, bilateral myoclonic jerks associated with rapid spike bursts on the EEG were observed and frequent errors were noticed in counting (Fig. 1).

Other EEG findings

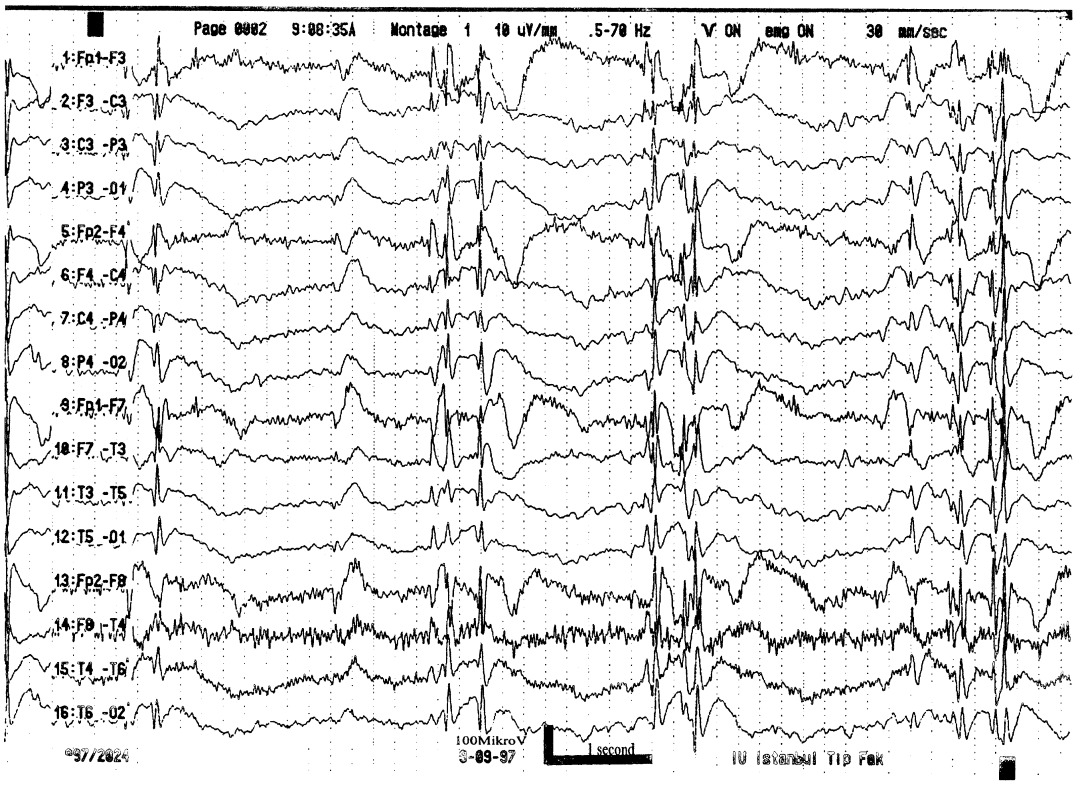
The background activity was normal in the interictal period, in all of the patients except one (case 3). In this latter case, mild slowing was attributed to high antiepileptic drug levels in two subsequent recordings. Hyperventilation procedure caused an increase in the epileptiform activity in three of the patients and photic stimulation disclosed prominent increases in the discharges in two cases with prominent eyelid myoclonia. Typical generalised spike and wave discharges following eye closure as well as photoparoxysmal responses were observed in the interictal EEGs of the cases 1 and 7 with eyelid myoclonia with absences.

Video-EEG examinations could be done in five patients in the interictal state. In two of those investigations, (cases 3 and 8) no seizures were observed; however, rare generalised spike and wave discharges were noted. In video-EEG studies of our previously published case 6, three absence seizures were recorded with marked perioral myoclonic movements almost synchronous with the spike component of the spike-wave discharges[12]. Absence seizures with eyelid myoclonia appearing on eye closure were observed in case 7. In case 5, phantom absences were demonstrable on video-EEG.

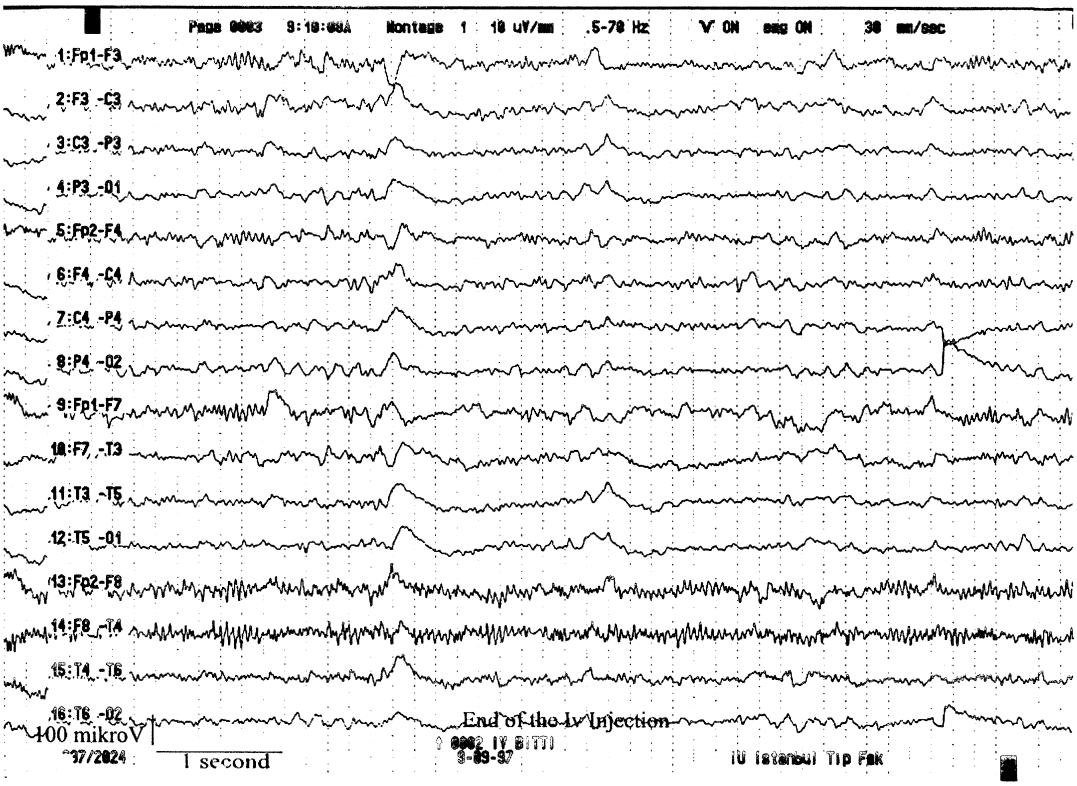
Syndrome diagnosis and correlation with the EEG features

With regard to the described clinical and EEG features of the patients, we attempted to classify them according to the known epileptic syndromes[11].

Two of them had the syndrome of juvenile myoclonic epilepsy (JME) (cases 2 and 3). The EEGs during ASE of these two patients had marked similarities. We observed rapid generalised spike (6 Hz) bursts associated with generalised myoclonus, followed by less prominent delta waves and frequent interruptions lasting 1–4 s during the ASE (Fig. 1). The main characteristics of the EEG pattern did not change through the recorded periods. Although these two JME patients showed a highly similar pattern of

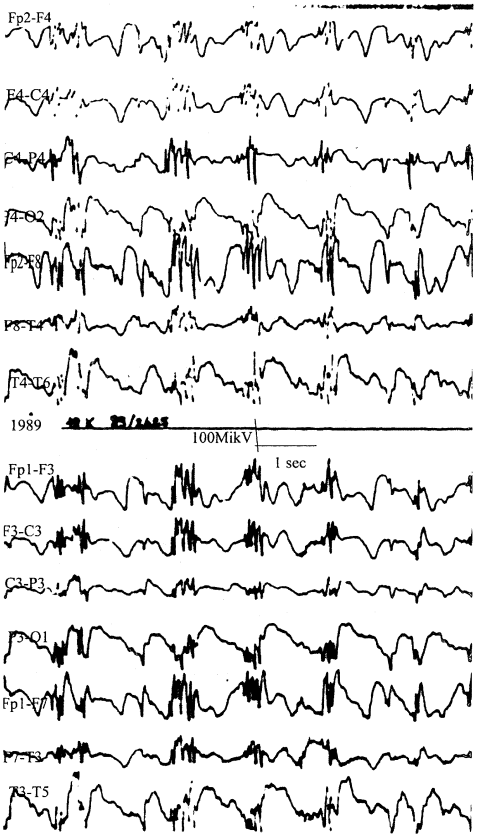


(*a*)

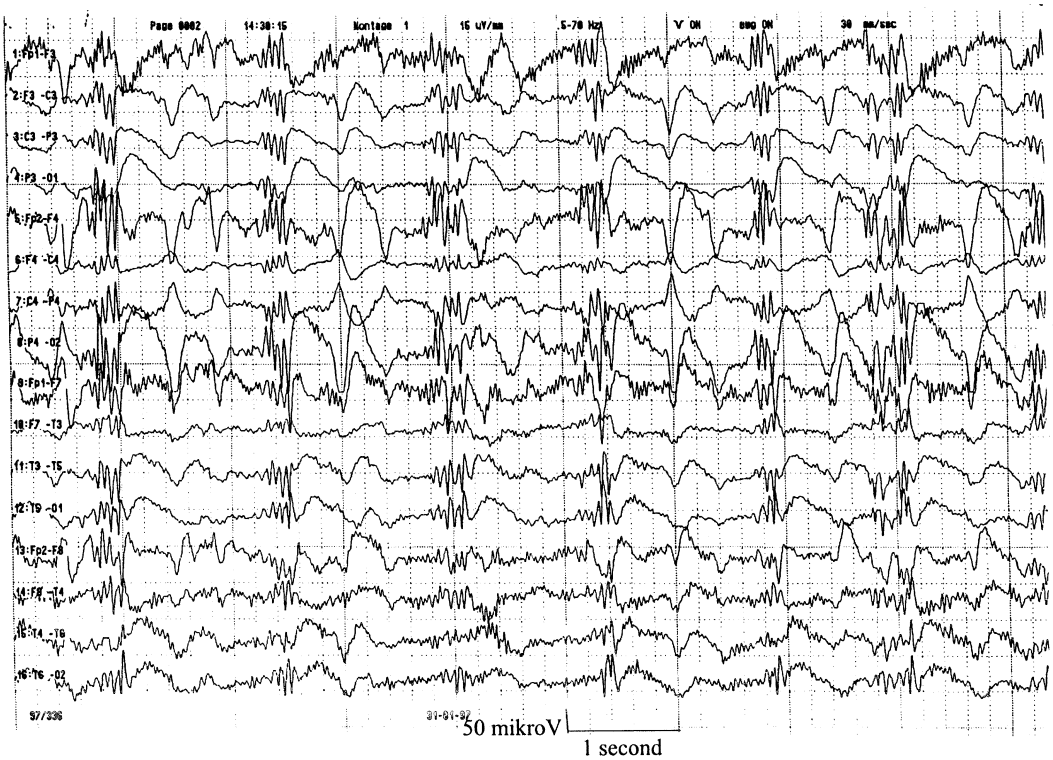


(*b*)

Fig. 1: (*a*) Video-EEG examples of case 3 with JME during ASE. Note that the discharges are discontinuous and non-rhythmic with prominent rapid (5–6 Hz) spikes associated with myoclonic jerks of the upper extremities and moderate confusion. The spike bursts are usually followed by delta waves and there are interruptions lasting a few seconds between the seizure discharges; (*b*) demonstrates the dramatic effect of IV clonazepam. The patient feels improvement, he falls asleep and the seizure discharges disappear completely.



(*a*)



(*b*)

Fig. 2: (*a*),(*b*): EEG tracings of case 4 with phantom absences and rare generalised convulsions showing the identical EEG pattern during 2 ASE attacks in the years 1989 (*a*) and 1997 (*b*). Note that the discharge is continuous with poly-spikes and prominent slow waves.

Table 2: EEG findings of the recurrent ASE cases.

	1	2	3	4	5	6	7	8
During ASE			Figure 1(a), (b)	Figure 2(a), (b)				
Morphology	Multi-SW	Multi-SW	Multi-SW	Multi-SW	Multi-SW	Multi-SW	Multi-SW	Multi-SW
Regularity	Yes	No	No	Yes	No	Yes	No	No
Continuity (interruptions)	Yes	No (1–3 s)	No (2–4 s)	Yes	No (2–4 s)	No (1–2 s)	No (1–6 s)	Yes
IV (CLZ)	Effective	Effective	Effective	Effective	Effective	Effective	Effective	Effective
Video findings	Not done	Not done	Difficulty in counting and frequent myo.	Not done	Mild confusion, difficulty in counting	Not done	Not done	Marked confusion
Observation of technician or physician	Eyelid myoclonus	Generalised myoclonus	Generalised myoclonus	Random jerks and rare eye blinking	Rare eyelid blinking	Perioral myoclonus	Eyelid myoclonus	Facial and generalised myoclonus
Interictal period								
Background activity	Normal	Normal	Mild slowing due to drugs	Normal	Normal	Normal	Normal	Normal
Routine EEG	SW, Right temporal S	N (Under VPA therapy)	Multi-SW	Multi-SW	N/rare Multi-SW	Rare SW (R > L)	Frequent SW	Rare irregular SW
Video-EEG findings	Not done	Not done	Multi-SW	Not done	Phantom absence seizures	Perioral myoclonia with absence seizures	Eyelid myoclonia with absence seizures	Normal sleep and wakefulness with rare SW on awakening
Eye closure	+ (SW)	N	N	N	N	N	+ (SW)	−
Hyperventilation	+	N	+/−	+	−	Mild +	−	−
Photic stim.	+ (15 Hz)	N	N	N	−	−	+ (on eye closure)	−

SW: spike and wave; CLZ: clonazepam; +: epileptic discharges started or prominently enhanced. R: right; L: left; VPA: valproate.

ASE on the EEG, their clinical courses were different regarding the response to AED therapy. Case 2 had only three attacks of ASE before appropriate treatment was started whereas the other patient (case 3) had frequent attacks despite treatment. On the other hand, in that latter case, ASE attack responded markedly to IV Clonazepam, like all of the other cases (Fig. 1(b)).

The youngest patient (case 8) had childhood absence epilepsy (CAE) with very frequent ASE attacks on awakening during childhood. However, the EEG pattern during the late phases of ASE of this case seemed to be somewhat atypical for this syndrome, regarding the marked irregularity. This case is still in remission for the past three years, but his long-term video-EEG demonstrates rare generalised spike and wave discharges in the awakening hours.

The remaining five patients with recurrent ASE could not be classified; they had no definite place in the existing classification of the IGE phenomena. Among these five cases, two cases had eyelid myoclonia with absence seizures (EMA) (cases 1 and 7). They both had only three to six ASE attacks before treatment, while they did not have any attacks after treatment. Their EEGs during ASE showed discontinuous repetitive discharges of multispikes and slow waves associated with eyelid myoclonia and impairment of consciousness. Both of these patients also had eyelid myoclonia with absence seizures of short duration appearing on eye closure and photoparoxysmal responses.

Two other patients with more numerous ASE recurrences had phantom absences (PA) and late onset generalised convulsions (cases 4 and 5). Their EEG patterns were not similar regarding continuity but they shared a similar multi-spike pattern. The patterns of the ASE EEGs did not change during the course over years (Fig. 2(a), (b)) in these two cases.

The remaining patient had perioral myoclonia with absences (PMA) (case 6). His EEG during ASE disclosed a highly regular pattern of 4 Hz spike-waves with 2–3 spikes per wave. There were rare and very short normal intervals[12].

Response to treatment and outcome

Since the effects of different antiepileptic drugs cannot be properly evaluated in such a small group, evaluation of the treatment effects is beyond the scope of this study. The follow-up period ranged from 2 to 12 years. Two patients were seizure-free after treatment with ethosuximide and clonazepam. Five patients were treated with low to moderate doses of valproate. Clonazepam was also effective in two patients. Carbamazepine was reported to worsen the ASE attacks in case 6. Ethosuximide was also used in three cases with absence seizures. The oldest patient (case 5) with a diagnostic delay of 40 years refused to take the medicine regularly. She used small doses of clonazepam, when her attacks recurred. Case 3 continues to have generalised tonic–clonic seizures and ASE attacks despite treatment with various combinations of appropriate antiepileptic drugs.

DISCUSSION

Nonconvulsive status epilepticus (NCSE) accounts for approximately one-quarter of all cases of status epilepticus[13]. The actual proportion may be higher because patients with NCSE often go unrecognised, or are mistaken to have behavioural or psychiatric disorders. The heterogeneous clinical presentation of NCSE indicates that EEG and a therapeutic trial of AEDs afford the best diagnostic measures in acute confusional states[2]. This disorder may be divided into generalised (absence) or partial (complex partial) forms. ASE is considered more frequent and is characterised by a continuous neurocognitive alteration[13]. ASE is reported to occur in 3% of all patients who have absence seizures[8]. Tomson *et al.*[14] reported the annual incidence of NCSE as 1.5 in 100 000 inhabitants. In their series, which included all NCSE cases, the median age at onset of status was 51 years, markedly older when compared with our group comprising only recurrent ASE cases[14].

For ASE to be distinguished from the state of focal NCSE (complex partial nonconvulsive status epilepticus) clinical and EEG findings should be evaluated together with extreme care and this sometimes causes great difficulty[1,15]. However, it is important to differentiate between complex partial status epilepticus (CPSE) and ASE since, in the case of ASE first choice treatment should include anti-absence drugs. Kaplan[16] evaluated causes of diagnostic and management delay and examined frequent diagnostic features in a retrospective study of 23 patients with NCSE. We also observed that the correct diagnosis was significantly delayed in the majority of cases.

Brinciotti *et al.*[17] observed an increased incidence of convulsions among the relatives of probands with NCSE, but no difference was noted between epileptic children with or without NCSE. We also noticed a remarkable family history in our small group.

Most of the cases had a history of epilepsy before ASE, although the first epileptic attack might be seen as an ASE in some of the relevant reports[2,10]. Approximately one-third of the NCSE patients had status as their first epileptic manifestation in the study of Tomson *et al.*[14]. Similarly, in our series, more than half of the cases had a history of epilepsy before ASE.

EEG is the best diagnostic tool for ASE. Except for rarely reported special findings such as generalised fast activity[18], the diagnosis becomes certain with the occurrence of continuous or frequently recurring generalised spike and wave discharges[8,19]. IV Clonazepam led to rapid resolution of ASE. Kaplan[16] reported the absence of evident epileptic findings on EEGs of some patients in a heterogeneous group of NCSE cases. In contrast, the number of spikes per wave was greater than one in our group of recurrent ASE cases. Granner and Lee[19] stated that the ictal discharges tended to be relatively slow in NCSE. We found also in some of the ASE EEGs, the frequency of the generalised spike and wave discharges slowed down to 1.5 Hz.

Inoue *et al.*[20] described a distinct epileptic syndrome associated with ring chromosome 20, with frequent seizures consisting of a prolonged confusional state and an ictal EEG pattern of bilateral high-voltage slow waves with occasional spikes. We investigated the suspected cases in our group, but could not find any case with this peculiar chromosomal abnormality.

Patients with JME can develop NCSE, which may be overlooked because of the subtle clinical symptoms[21]. The EEG pattern of ASE is highly typical with rapid spike bursts associated with myoclonus in our two JME cases.

It is reported that in some cases ASE was frequently recurring and seen as a major seizure type[8,9,14]. Guberman *et al.*[1] mentioned recurring attacks in spite of antiepileptic drug treatment. Berkovic *et al.*[9] showed that VPA prevented recurrence of ASE in the group with unclassified IGE. Despite the limited number of the cases, our study suggests that ASE frequently recurs in some cases and can be the characteristic seizure type for some epileptic patients.

Recently new epilepsy syndromes frequently seen in ASE are being reported. Among those, epilepsy with eyelid myoclonia and absences (EMA), which

was seen in two of our cases, was the first one[22]. Wakomoto *et al.*[23] described another case with EMA in whom ASE developed shortly after awakening.

It has been reported that ASE appears to be related with the highest prevalence to the syndromes of PA with generalised tonic–clonic seizures and PMA[7]. It is possible to diagnose these syndromes with video-EEG recordings. From this point of view, it is necessary to examine the NCSE cases with video-EEG recordings, and a long follow-up period is required. Our study confirmed that the syndrome with mild (phantom) absences and late onset generalised convulsions is associated with recurrent ASE[24]. Our long-term follow-up demonstrated that the typical EEG pattern did not change during the course despite treatment in this peculiar syndrome. PMA is another rare and recently described epileptic state with distinctive clinical features[25,26].

It has been reported that NCSE responded to treatment with carbamazepine, with increased intracranial pressure and transient abnormalities observed on MRI[27]. One of our patients was markedly worsened when treated with carbamazepine because of a wrong diagnosis of partial epilepsy. Intravenous valproate may provide an effective treatment alternative to benzodiazepines in ASE without their associated morbidity[28].

There is no convincing evidence of any brain damage related to recurrent ASE in our study. Our study showed that recurrent ASE permitted a normal life span for most of the patients and supported the opinion that nonconvulsive status epilepticus is not very harmful[3,4]. Krumholz[29] reported that typical ASE does not appear to have very serious consequences and may be a type of 'inhibitory' seizure.

It is remarkable that ASE can recur in some patients and most of them could not be classified according to the known epileptic syndromes. The recurrent type of ASE may provide an opportunity for a better understanding of epileptogenesis and the mechanism of status epilepticus in future studies.

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