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Case report

Idiopathic late-onset absence status epilepticus: A case report with an electroclinical 14 years follow-up

Stefano Pro *, Edoardo Vicenzini, Franco Randi, Patrizia Pulitano, Oriano Mecarelli

Department of Neurological Sciences, Neurophysiopathology Unit, Sapienza University of Rome, Viale Regina Elena 336, 00185 Rome, Italy

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ABSTRACT

Late-onset absence status epilepticus (ASE) may be observed in adult and elderly patients as a late complication of idiopathic generalized epilepsy or *de novo*, usually related to benzodiazepines withdrawal, alcohol intoxication or psychotropic drugs initiation, but without history of epilepsy. EEG may be highly heterogeneous, varying from the 3 to 3.5 Hz spike-wave discharges typical of idiopathic generalized epilepsy to asymmetric irregular sharp and slow wave complexes. We report the clinical and neurophysiologic 14 years follow-up of a now 86 years-old woman, in whom we observed – at the age of 72 – an idiopathic late-onset ASE, with a good clinical response to lamotrigine monotherapy, but with the persistence over years of the same interictal 3–3.5 Hz spike-wave epileptic activity at EEG. This case is singular because, with the available long follow-up, indicates that idiopathic generalized epilepsy may also occur in the elderly, with a late-onset ASE presentation. In this condition, it is particularly important to underline the essential role of EEG (urgent and ambulatory) for the diagnosis, management and monitoring of the disease.

1. Introduction

Late-onset absence status epilepticus (ASE) may occur in adult and elderly subjects as a late relapse of idiopathic generalized epilepsy, which had remitted after puberty or juvenile age, without any identifiable triggering factors,[1–3] or in patients *de novo*, without any seizure history. The *de novo* late-onset ASE has been more frequently described in women, usually provoked by the benzodiazepines withdrawal, alcohol intoxication or psychotropic drugs initiation.[4–7]

In the late-onset ASE clinical symptomatology is characterized by a prolonged and fluctuating confusional state with or without minor abnormal motor component.[8,9] EEG presents several kinds of ictal pattern, varying from typical or atypical spike-wave discharges to asymmetric irregular sharp and slow wave complexes with anterior predominance.[6,8] Furthermore, interictal EEG is usually irregular as regards to the background activity organization. In these cases, clinical and EEG normalization is obtained with intravenous benzodiazepine or phenobarbital administration, without the relapse of the epileptic activity. As a matter of fact, long-term treatment is generally not required in *de novo* late onset ASE, if provoking factors can be avoided.[4,5] Emergent EEG is essential in these cases to confirm the clinical diagnosis of non-convulsive status epilepticus (NCSE).

We report the singular case of a long clinical and neurophysiologic 14 years follow-up of a now 86 years-old woman, in whom we diagnosed an idiopathic late-onset ASE at the age of 72, with a good clinical response to lamotrigine monotherapy, but with the persistence over years of the same interictal 3–3.5 Hz spike-wave epileptic activity at EEG. This case shows that idiopathic generalized epilepsy may also occur in the elderly, with a late-onset ASE presentation and that EEG has an essential role in the diagnosis and management.

2. Case description

In February 1995, a 72 years-old woman was admitted to the emergency room of our hospital for the acute onset of an altered state of consciousness, completely and spontaneously recovered after 5 h. In her past medical history, similar episodes had already presented since the age of 54, the first occurring in 1977. These episodes were characterized by acute onset of mental confusion and ideomotor slowing, apraxia and speech arrest, without motor abnormalities or sphincter release, usually showing full regression in less than 5–6 h and frequently occurring in the mornings at awakening, with a frequency of 2/3 per year. Being transient and with full spontaneous recovery, the patient underestimated these symptoms and she did not refer to any medical control or treatment. Moreover, from the interview at admission, she did not reveal any other relevant illness and there was no evidence of benzodiazepines or psychotropic drugs intake.

* Corresponding author. Tel.: +39 06 49912875; fax: +39 06 49912657.
E-mail address: stefanopro1974@libero.it (S. Pro).

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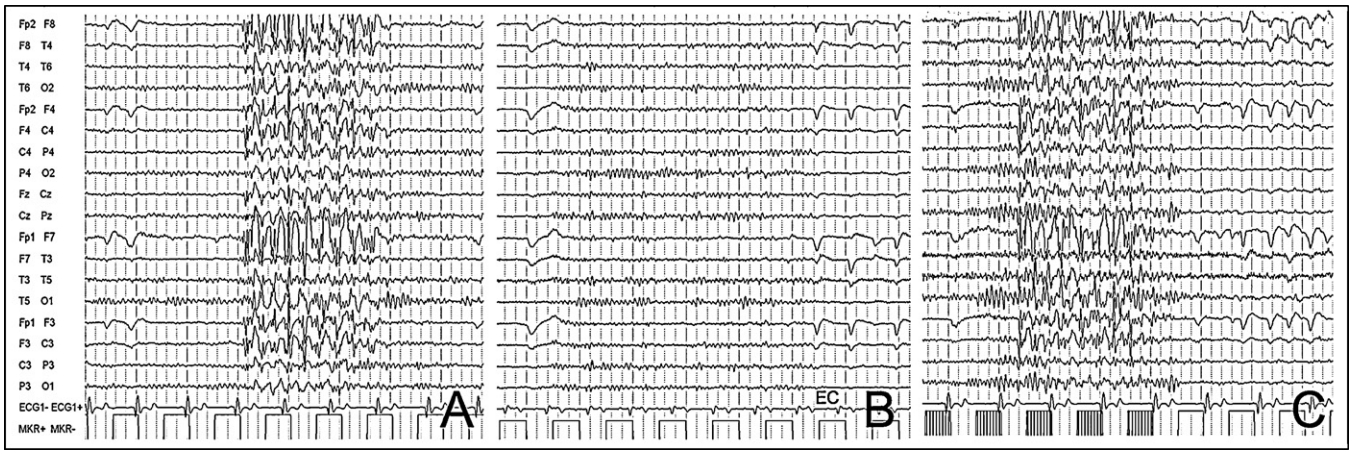

Fig. 1. Standard EEG (S/EEG) at first admission in hospital at the age of 72 years. (A) Generalized spike- and polispike-wave discharges at 3–3.5 Hz of higher voltage in the anterior regions; (B) occipital alpha rhythm with blocking response to eye opening (EO); (C) photoparoxysmal response at intermittent photic stimulation at 18 Hz. High Filters: 30 Hz; Low Filters 0.1 Hz; Notch Filter 50 Hz; Sensitivity: 100 μV/cm; speed: 30 mm/s.

Neurological examination at admission was normal, patient was awake and collaborative, and no focal neurological sign was observed. Routine blood exams, including glycemia, glicated hemoglobin and electrolytes, electrocardiogram and chest X-rays were normal. Cerebral Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) showed only slight subcortical brain atrophy with very mild brain ventricular dilatation, compatible with age.

Standard EEG (S/EEG) recording at the emergency room, when the patient had already returned asymptomatic, showed a regular and symmetrical 3–3.5 Hz generalized spike-wave and polispike-wave interictal discharges (Fig. 1A), with normal posterior alpha-rhythm blocked by eye opening (Fig. 1B), and photoparoxysmal response at intermittent photic stimulation (Fig. 1C). Duration of the epileptic discharges varied from 0.5 to 10 s. Patient was immediately submitted to EEG monitoring with ambulatory EEG (A/EEG) recording for 24 h to quantify the epileptiform abnormalities. The total number of epileptic discharges was 544/24 h, their total duration 1820.3 s and the mean discharges duration 3.3 ± 5.6 s. The epileptic discharges occurred mainly in the mornings, 2 h after awakening, they were of abrupt onset and termination and completely disappeared during sleep.

An idiopathic *de novo* late-onset SE without triggering factors was hypothesized and the patient was discharged with lamotrigine monotherapy, titrated up to final dosage of 200 mg/day. In the following year, the patient did not present any new clinical episode, even though 2 A/EEGs, performed 6 and 12 months from initial hospitalization, showed the persistence of a relevant number of epileptic discharges, but with a shorter duration (Table 1).

In the following 14 months, for the apparent clinical wellness, the patient arbitrary decided to reduce and suspend therapy for three times, each followed by the re-appearance of the confusional states. During the third episode she was brought again to our observation and, while symptoms were still ongoing (ideomotor slowing and speech blurring), S/EEG showed a status epilepticus with a continuous, diffuse, 3–4 Hz spike/polispike-waves epileptic activity, predominant in the frontal regions and without total suppression of background activity in the posterior regions (Fig. 2). Clinical symptoms and status epilepticus spontaneously disappeared few minutes after EEG initiation and the patient was discharged with add-on therapy: lamotrigine 200 mg/die, valproate 1000 mg/die.

Three months thereafter, she refused to carry on the treatment with valproate for the occurring of side effects (hair loss and subjective drowsiness) and she agreed to continue only lamotrigine at the dosage of 200 mg/die, but now with good compliance. For the following 12 years until nowadays she never presented again clinical episodes.

The follow-up is now arrived at 14 years, 12 years after the last symptomatic ASE, and S/EEGs and A/EEGs have been performed regularly, once a year, to monitor the epileptic activity. All these EEGs show the persistence of the epileptic activity, with the same morphological spike-wave pattern at 3–3.5 Hz, but with reduced duration and frequency of discharges; notwithstanding, in none of the EEGs a total disappearance of the epileptic activity had been observed. She is now in good state of health and she is 86 years-old, with a preserved cognitive/behavioral status as documented at the Neuropsychological testing, including Mini Mental State, Set test, Raven Spatial Recall test, Digit Span and Wechsler Memory Scale.

Interestingly, in her family history, she has a 16 years-old nephew, son of her daughter, that had been suffering from febrile seizures during the first year of life and that, at age 8, presented at home an episode of loss of consciousness that was not clearly interpreted as epileptic. Furthermore, several following EEG controls reported epileptiform abnormalities in the left central temporal regions during sleep and atypical diffuse paroxystic discharges at awake, but patient remained, up-to-now, asymptomatic and neuroimaging was normal.

Table 1
Ambulatory EEG 24 h (A/EEG) at first hospital admission and during the first 12 months follow-up. At month 6th and 12th epileptic discharges (EDs) were reduced only slightly in the total number, but with a shorter duration.

A/EEG	Total number of EDs	Total duration of EDs (s)	Mean duration of EDs (s)
Admission	544	1820.3	3.3 ± 5.6
6 months follow-up	422	589.2	1.4 ± 3.2
12 months follow-up	385	566.3	1.8 ± 2.8