

Absence status in the elderly as a late complication of idiopathic generalized epilepsies

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ABSTRACT – The main categories of nonconvulsive status epilepticus are: complex partial and absence status. Absence status was reported to occur *de novo* in later life as a situation-related, single event. We report four cases of absence status with presentation after the age of 60 years. At admission, no history of epilepsy had been mentioned. The preliminary diagnosis of absence status occurring *de novo* in later life had to be changed on completion of case histories. All patients had suffered from idiopathic generalized epilepsy with absence seizures, which had resolved after puberty. A second peak of IGE, with repeated episodes of absence status after the menopause and without identifiable triggering factors is assumed.

Key words: nonconvulsive status epilepticus, absence status, epilepsy, elderly, idiopathic generalized epilepsy

Nonconvulsive status epilepticus (NCSE) can be classified as complex or simple, partial status epilepticus (CPSE and SPSE) as well as absence status (AS). NCSE in the elderly mostly presents as CPSE (Shet *et al.* 2006, Tomson *et al.* 1992), whereas AS in later life is often regarded as a situation-related single event (Ellis and Lee 1978, Schwartz and Scott 1971, Thomas 1999, Thomas and Andermann 1994). However, Porter and Penry (Porter and Penry 1983) suggested that AS is rarely seen in patients without pre-existing epilepsy, even if AS occurs in later life.

We report the case histories of four patients from a large outpatient seizure clinic, to support the view that AS in the elderly may occur as a late

complication of idiopathic generalized epilepsy (IGE) after a long, seemingly seizure-free period.

Case reports

Patient 1

A 60-year-old lady was admitted disoriented and unable to follow a simple conversation. On the basis of the EEG, an AS was diagnosed and successfully treated with intravenous diazepam. The patient did not report any paroxysmal events in the past. A diagnosis of AS occurring *de novo* in later life was made. There was no history of alcohol abuse or benzodiazepine intake. Vascular encephalopathy, diagnosed on the basis of a history of

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hypertension and a mild cortical atrophy documented on CT-scan, was considered as precipitating factors for the occurrence of AS. With repeated examinations after a relapse of AS, careful history-taking revealed a pre-existing, childhood absence epilepsy (CAE) since the age of six years. These absence seizures stopped spontaneously at the age of 14 years without any antiepileptic drug treatment, but during the following years a few generalized tonic-clonic seizures (GTCS) occurred. The patient did not seek any medical attention at that time and she concealed all of her seizures from her family and friends because she felt ashamed. After appropriate counseling and introduction of long-term antiepileptic drug (AED) therapy consisting of valproic acid (VPA), the patient became seizure-free.

Patient 2

A 64-year-old lady was admitted with speaking difficulties and behavioral abnormalities. At admission, no lateralizing neurological abnormalities were found. Past medical history, particularly drug history was unremarkable. CT-scan and MRI revealed mild, diffuse cortical atrophy. The CT-scan was repeated four years later with unchanged results (figure 1). The EEG (figure 2) exhibited continuously occurring, bilateral rhythmical spikes and waves,

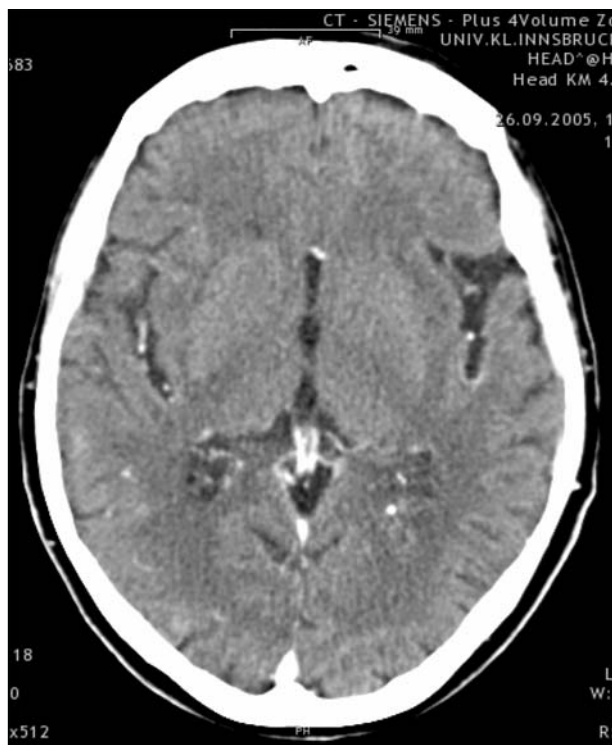


Figure 1. Patient 2, female, CT-scan performed four years after first presentation. Mild diffuse cortical atrophy and leucencephalopathy, unchanged in comparison to CT performed at the time of absence status.

and *de novo* AS in later life was diagnosed. AS stopped after treatment with iv diazepam. The EEG at follow-up showed repeated bursts of bilateral spikes and waves. The patient then reported daily lapses of consciousness during her school days. These absence seizures stopped at the age of 14 years, spontaneously, without treatment. At age of 55 years, she had had some GTCS and had experienced several prolonged episodes of changed behavior similar to the AS leading to admission. Treatment with VPA was started and the patient remained free of any attacks.

Patient 3

A 64-year-old lady was admitted with speech problems and difficulties in daily activities. The symptoms had occurred suddenly, and a CT-scan revealed a mild, diffuse, cortical atrophy accentuated over the left hemisphere, and a transient ischemic attack in the left medial, cerebral artery was assumed. Previous medical and drug history were uneventful and the symptoms resolved within one day. She was discharged from hospital with low dose aspirin and statins. After a further episode, the patient was admitted to our outpatient clinic where an EEG was performed which showed bilateral, 2-3/sec rhythmical spikes and waves and an AS was diagnosed. Clinically, the patient was grossly disoriented, and subtle twitches of facial muscles were observed. After 4 mg lorazepam intravenously, both the EEG and the patient's condition normalized. Periods of abnormal behavior had been observed since the age of 55 years, with feelings of general floppiness, inability to put on her clothes, disorientation, and inappropriate actions in her daily life. Furthermore, she reported weekly absence seizures since her "young days". A diagnosis of a generalized seizure disorder was established. The patient was put on valproic acid and no further episodes of NCSE occurred.

Patient 4

A 78-year-old lady slipped into a confusional state for several hours, the morning following a night with profound sleep deprivation. The confusional state was punctuated by a GTCS, which led to admission to the hospital. Except for slow mentation no other neurological abnormalities could be found. A brain CT scan showed mild, diffuse, brain atrophy and the EEG exhibited generalized spikes. A diagnosis of AS occurring *de novo* in later life was made. As there was no history of regular benzodiazepine or alcohol intake, her severe sleep deprivation and the diffuse brain atrophy were considered as triggering factors. At follow-up, a striking physiognomonic similarity to patient 1 was noticed. She was her sister. She then revealed the typical medical history of a CAE complicated by GTCS at the age of 54 and 78 years, and repeated episodes highly likely to be AS since her fifties. The patient was put on valproic acid and no further seizures occurred.

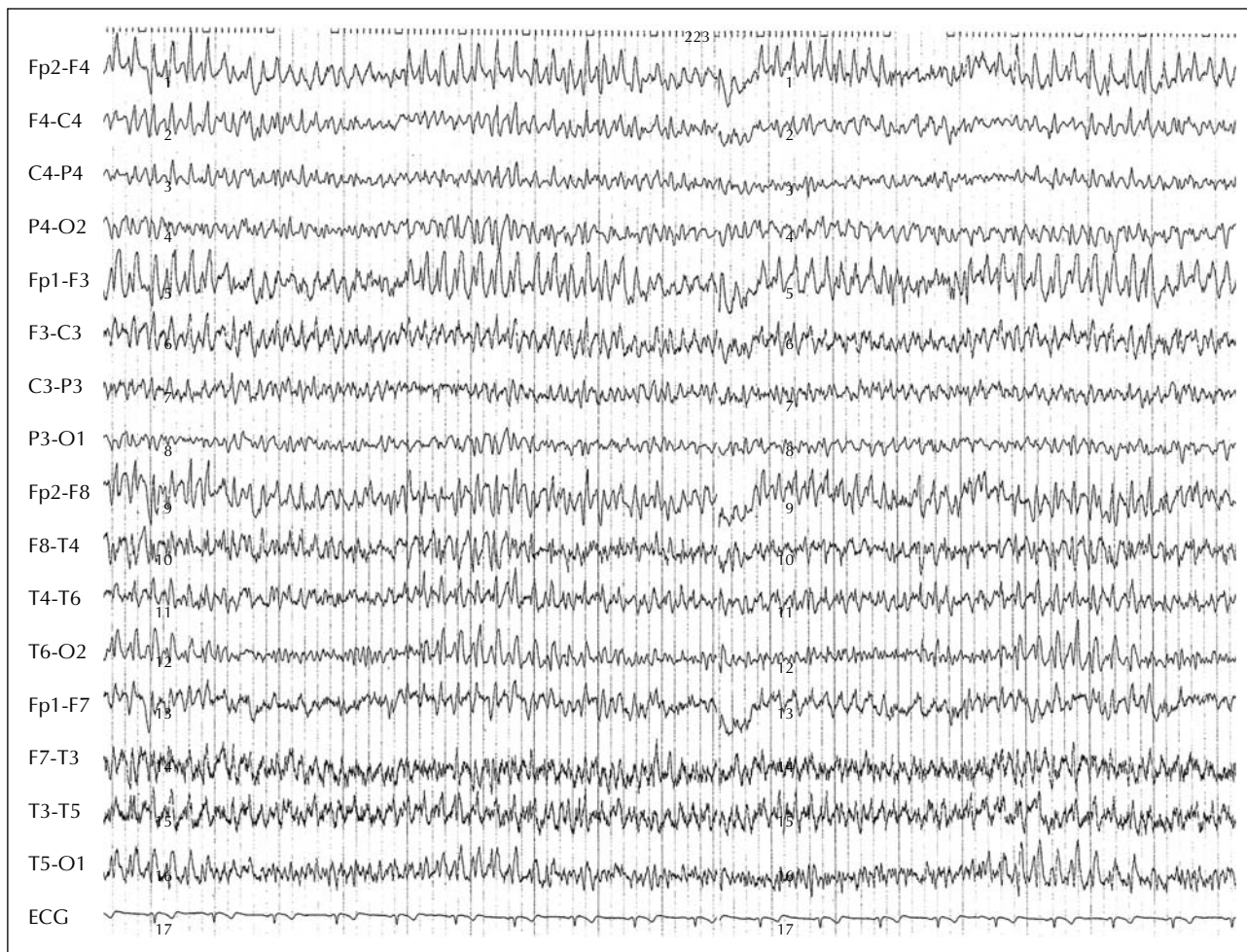


Figure 2. Patient 2, female, 64 years. tc 1.0 F 70. EEG during absence status. Continuous 3-4/sec spikes and waves maximal over the anterior regions. Note the intermittently occurring, 7-8/sec basic rhythm over the posterior regions.

Discussion

All our patients share some striking similarities: 1) age at clinical presentation of AS between 60 and 78 years; 2) presence of mild cortical atrophy on CT or MRI and 3) denial of a history of seizures on admission.

None of our patients had a history of alcohol abuse or consumption of benzodiazepines, therefore, we initially assumed an AS occurring *de novo* in later life with mild vascular encephalopathy as a precipitating factor. Without a close follow-up this would have been the final diagnosis. After additions to the historical data, a pre-existing IGE was obvious. A marked reduction in seizure frequency had occurred after puberty in all four patients. However, rare GTCS and short lapses of consciousness scattered over the years indicated that the epilepsy was not cured. Notably, the patients had not taken any antiepileptic medication previously, suggesting a mild course for the disease. With appropriate AED treatment (all patients were treated

with sodium valproate) after the presenting AS, the seizure disorder was well controlled in all patients.

Triggering factors for situation-related AS in the elderly were protean, including psychotropic and other seizure-precipitating drugs such as aminophylline, benzodiazepine withdrawal, metabolic imbalance, systemic infections and fever, alcoholism and dehydration (Thomas and Andermann 1994). However, none of our patients had any of the aforementioned precipitating factors for the AS. Although cerebrovascular disease is the most common cause of acute symptomatic seizures and remote symptomatic epilepsies in the elderly with predominating partial seizures (Tallis *et al.* 2002), absence seizures are not reported in this context (McBride *et al.* 2002). Beyond that, none of our patients suffered from significant cerebrovascular or neurodegenerative disease and the mild diffuse cerebral atrophy found on CT (*figure 1*) or MRI was regarded as cryptogenous, which is a frequent finding in late-onset epilepsies (Regesta and Tangelli 1992).

A case control study on 228 patients with late-onset epilepsy recognized that cryptogenous cerebral atrophy (CCA) is significantly more often associated with generalized seizures and a positive family history for epilepsy than patients with symptomatic epilepsies secondary to diffuse cerebral atrophy due to an identifiable cause, suggesting a strong genetic background in the cryptogenous group (Regesta and Tangelli 1992). Therefore, in late-onset epilepsies, CCA *per se* can not be taken as an etiologic factor, and does not exclude an IGE.

Persons with epilepsy, especially elderly, tend to deny their history of seizures or do not report it immediately. The reason for this underreporting might be fear of social consequences, of losing their driving license, the commonly held belief in social stigmatization (Spatt *et al.* 2005) or they consider their fits not to be epileptic. Patients with absence seizures during their school days might have forgotten them when asked after years (recall bias) or they may not even have been diagnosed properly at that time (some of our patients had their absences during their childhood in the 1930s and 1940s). This bias may lead to a notorious underestimation of previous epilepsy in retrospective series, epidemiological studies and genetic analyses of families with epilepsy, as well as previously published case series of IGE with late onset (Luef *et al.* 1996, Marini *et al.* 2003).

In contrast to situation-related cases, our patients suffered repeated episodes of NCSE with deterioration later in life, indicating the need for appropriate AED treatment. However, all patients had a previously benign course, with many years of seizure-freedom without any AED treatment. The cause of a second peak of IGE in older age groups remains an enigma (Trinka 2005). Interestingly, all reported patients in our series were women. The female preponderance is consistent with previous reports on AS in the elderly (Gastaut 1981, Thomas and Andermann 1994). Gastaut (Gastaut 1981) explained both the female preponderance and the age after menopause to be the result of hormonal influences.

Although this is an anecdotal report of four patients observed over the years, it supports the view that absence status in the elderly is not always a situation-related condition, but may occur as a late complication of an otherwise benign idiopathic generalized epilepsy. □

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