A case of systemic lupus erythematosus with complex partial status epilepticus

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ABSTRACT – We describe a woman with systemic lupus erythematosus who had complex partial status epilepticus as the sole epileptic manifestation. The patient presented with a confusional state, initially considered to be psychotic in nature, due to SLE. However, the EEG demonstrated the epileptic etiology of her symptoms. In patients with lupus presenting with prolonged episodes of confusion and behavioural disturbance, an EEG should be performed, whenever possible during the episode, even in the absence of a previous history of epilepsy.

Keywords: systemic lupus erythematosus, complex partial status epilepticus, confusional state, non-convulsive status, psychosis

Systemic lupus erythematosus (SLE) is frequently associated with seizures (Palace and Lang 2000), which can be partial or generalized. Patients may also experience status epilepticus, rarely presenting as complex partial status epilepticus (CPSE). Our patient was initially considered to be suffering from SLE with psychosis, but the symptoms were later attributed to CPSE. This case report describes SLE presenting uniquely as CPSE.

Case study

In late October 2000, a 56-year-old, right-handed female with a 33-year history of SLE and under treatment with immunosuppressive drugs and steroids, was admitted to our hospital for a first episode of confusional state preceded by nausea. Neurological examination was normal. Brain CT scan revealed diffuse cerebral atrophy. Magnetic resonance imaging (MRI) of the brain also disclosed diffuse cerebral atrophy, mainly in frontal and temporal lobes, and some lacunae lesions in the white matter and basal ganglia.

Her confusional state disappeared the next day without any medication apart from antibiotics and vitamins. After the first episode, when she was no longer confused, an electroencephalogram (EEG) showed a 9-10 Hz background activity and occasional, diffuse slow (5-7 Hz) wave bursts with a frontal maximum. Carbamazepine (CBZ) was then administered at a daily dose of 200 mg in view of the possibility that her confusional state was an epileptic event. One month later, she again had an episode of confusion and abnormal
behaviour, preceded by vomiting, during which she complained of a feeling of heaviness in her legs. She was agitated in bed and said “I want to have an easy death.” This episode involved no automatisms or clinical manifestations. At this time, since her neurological examination was normal and her symptomatology was considered SLE-related, we thought that she had some type of psychosis.

An EEG showed 150 µV, diffuse 5-7 Hz continuous slow waves (figure 1). The EEG and clinical symptoms improved following intravenous administration of diazepam (DZP). She was able to speak normally, her mood improved and psychomotor manifestations disappeared. However, her confusional state reappeared. An intravenous drip of phenytoin (PHT) was then given, and her confusional state disappeared within three days. The patient was disoriented during these episodes and retained little memory of them. No abnormal metabolic data were subsequently found. A diagnosis of non-convulsive status epilepticus (NCSE) was made and treatment with PHT was continued.

During the next three years she experienced a total of seven episodes of CPSE. Nausea or vomiting preceded all but one of the CPSE episodes, each of which continued for two to six days. The ictal EEG of the fourth episode showed high voltage, 2.5-3 Hz, continuous slow waves or 2.5-3 Hz, diffuse spike and wave complexes, which partially formed triphasic-like waves. Ictal EEGs of other episodes also showed continuous high voltage 2.5-3 Hz, slow waves or diffuse, continuous, 2.5-3 Hz spike and wave complexes. No correlation was found between these confusional episodes and serological evidence of worsening SLE. Steroids could not be incriminated as she was on this regimen only when she experienced the first two episodes, but were discontinued and only immunosuppressive drugs were used thereafter.

During the fourth episode, administration of intravenous diazepam modified the continuous diffuse high-voltage, 2.5-3 Hz slow waves over the frontopolar regions. The interictal EEG sometimes also demonstrated 2.5-3 Hz high-voltage slow waves over the left frontal areas. We then considered that her epilepsy was probably frontal in origin.

With evolution, interictal, abnormal slow waves were more frequently recorded. Recently, delusional symptoms were observed during the transitional period following episodes of nonconvulsive status. For example, after her seventh episode she cried, saying “My husband and daughter were hurt in a traffic accident!”. Brain atrophy at CT was seen to be progressing gradually.

Moreover, between the fourth and fifth episode of NCSE, she experienced a wandering episode. Although we were not able to obtain detailed information on this episode we

![Figure 1. Ictal EEG of a second episode of CPSE. High-voltage, 5-7 Hz, continuous slow waves are diffused. The patient had never experienced other episodes evocative of partial seizures.](image-url)
believe that, because it lasted for a few hours, this was not
a complex partial seizure but yet another episode of NCSE.
The patient never experienced other types of seizures or
manifestations that could be considered as being of an
epileptic nature (staring, automatisms or other focal mani-
festations). After her fourth episode, phenytoin was dis-
continued and replaced by sodium valproate. Currently
she receives a combination of sodium valproate and car-
bamazepine. Although blood levels of these drugs are
within the usual range, she still experiences episodes of
NCSE.

Discussion

We describe the clinical, electroencephalographic and
neuroimaging findings of a woman with SLE who initially
showed NCSE and which later resolved to the frontal lobe,
after IV benzodiazepines revealed it to be CPSE.
We were at first uncertain whether the continuous slow
waves showing partially triphasic wave on the EEG were
ictal, due to a disturbance of consciousness or psychosis.
However, the slow waves disappeared, her level of con-
sciousness improved following intravenous injection of
DZP, and we could find no abnormal metabolic data.
Fountain and Waldman (2001) reported that benzodiaze-
pine administration may help distinguish the sharp waves
of epileptic origin from those resulting from metabolic
encephalopathy. They found that when a benzodiazepine
induces a resolution of electrographic changes and clin-
ic improvement, the epileptic origin becomes more evi-
dent. In short, because a resolution of electrographic
changes and clinical improvement was achieved by diaz-
epam in our case, the confusion was not considered as an
encephalopathy. Besides, rhythmic bursts, such as those
we recorded, are not usually seen in encephalopathic
processes. Psychosis was also eliminated for the reasons
described above.
On the ictal EEGs of this case, clearly lateralized epilep-
tiform discharges were not recorded, although frontal dis-
charges were eventually seen; however, this may be due to
the fact that ictal onset was not recorded on the scalp EEG.
This is also supported by the fact that the interictal EEG
sometimes showed slow waves predominantly over the
left frontal area and ictal, continuous, diffuse high-voltage
slow waves over the frontal pole became evident following
administration of diazepam. Mayeux and Lüders
(1978) suggested that a prolonged complex partial seizure
resulting in CPSE should have continuous complex symp-
toms accompanied by a continuous electroencephalo-
graphic seizure pattern that is either focal or secondarily
generalized. They also suggested that an ictal EEG of CPSE
can take the form of generalized waves.
Both diffuse and focal electroencephalographic abnor-
malities have been reported in lupus (Koppel 1997). Our
case is thought to present the long-lasting periods of
confusion of CPSE reported by Gastaut and Tassinari
(1975). Some authors have observed CPSE occurring in
lupus. Fernández-Torre et al. (2003) reported the case of a
patient with SLE who experienced episodes of extratem-
poral CPSE, having also presented left, hemisporal, par-
tial motor seizures evolving occasionally into generalized
tonic-clonic attacks. They suggested that CPSE should be
considered as a possible cause of altered mental status in
lupus and that an EEG is essential to make an accurate
diagnosis. This becomes particularly true when consider-
ing the case of our patient who had never presented with
any other type of seizure suggestive of an epileptic origin
for her confusional attacks.
In conclusion, EEG should be conducted in all SLE patients
presenting with episodes of acute confusional state, even
in the absence of a history of epilepsy.

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