Status Psychosus epilepticus; A Special Type of Non-Convulsive Status Epilepticus
État de mal épileptique psychiatrique, un aspect inhabituel d'état de mal épileptique non-convulsif

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Abstract:
It has long been stated that psychotic behavior may be the only presenting symptom in psychomotor Epilepsy. Those epileptic equivalents last for a few minutes. This work is aiming at defining a special type of non convulsive status epilepticus, that can be called "Status Psychosus epilepticus" (SPE), which is defined as a Prolonged disturbed behavior due to disturbed brain electrical activity and characterized by:
(1) Psychotic behavior as the only presenting manifestations.
(2) Persistent for weeks (one to six weeks in the present study)
(3) Complete and persistent response of the Psychotic behavior, in these epileptic states, to antiepileptic drugs, alone.
Subjects and methods: Authors report 37 patients, admitted with psychotic manifestations for at least one week, and drug naïve. They were subjected to full clinical and psychological assessment, in addition to EEG. Results: MMSE range from 13 to 26. EEG showed periodic lateralized epileptiform activity. IV diazepam improved 62%. Monotherapy was effective in 75.7% of cases. 24% needed polytherapy. Conclusions: SPE may last for weeks. SPE can be misdiagnosed as brief psychotic disorder, or even as Schizophrenia; if clinical examination and EEG were not done for every suspicious case.
Keywords: Non-convulsive status epilepticus- Brief psychotic disorder- EEG- Schizophrenia.

Résumé:
Il a longtemps été dit que le comportement psychotique peut être le seul symptôme de l'épilepsie psychomotrice. Ces équivalents épileptiques ne durent que quelques minutes. Ce travail vise à définir un type particulier de l'état de mal non convulsif, qui peut être appelé «État de mal épileptique Psychotique» (EMP), qui est défini comme un comportement perturbé prolongée due à la perturbation de l'activité électrique du cerveau. Il se caractérise par:
(1) Comportement psychotique comme seule manifestation clinique.
(2) La persistance pendant des semaines (une à six semaines dans la présente étude)
(3) La réponse complète et persistante du comportement psychotique, dans ces états épileptiques aux médicaments antiépileptiques, seuls.
Sujets et méthodes: Les auteurs rapportent 37 patients, admis avec manifestations psychotiques pendant au moins une semaine, et jamais traités. Ils ont été soumis à une évaluation clinique et psychologique, en plus de l'EEG. Résultats: Le MMSE variait de 13 à 26. L'EEG a montré une activité épileptique latéralisé périodique. Le diazépam en intraveineux a apporté une amélioration dans 62%. La monothérapie était efficace dans 75,7% des cas et 24% avaient besoin d'une polythérapie. Conclusions: L'EMP peut durer des semaines et peut être diagnostiqué à tort comme un trouble psychotique bref, ou même comme une schizophrénie, si l'examen clinique et l'EEG ne sont pas demandés devant tout cas suspect.

Mots-clés: Etat de mal épileptique non-convulsif- Trouble psychotique bref- EEG- Schizophrénie.

Introduction:
In 1822, Prichard [1] described cases of epileptic fugue and furor as well as «epileptic ecstasy». Bright [2], as well as Charcot [3], described fugue states, and Hughlings Jackson [4] described such a condition in temporal lobe epilepsy.
In 1960, Lennox and Lennox [5] used the term petit mal status for psychiatric conditions associated with continuous bifrontal spike-wave activity and with duration of hours to days.
One of the authors who explore the relationship between temporal lobe pathology and psychosis was Wilhelm Sommer while working as a psychiatrist in the lunatic asylum of Allenberg in Germany. He wrote two extensive papers on these topics. In the first of these, Sommer [6] analyzed neuropathological findings in the Ammon's horn of 90 epilepsy patients. This material represented all previously published cases supplemented by five of Sommer's own patients from Allenberg. For each of the 90 patients Sommer specified seizure symptomatology, psychopathology and neuropathological findings.
Sommer described a characteristic cell loss in the CA-I Sector of his cases (Sommer's sector). In his opinion pathological changes in the hippocampus, which he thought was a major sensory center, were of significant etiological importance for the development of epilepsy and substantiated this by emphasizing the frequency of sensory impairments and hallucinations in epilepsy patients.
One year later, Sommer [7] published an article on post-epileptic insanity in which he developed an extensive classification of the various subforms of epileptic

insanity, including also etiology and prognosis. He based his hypotheses on quantitative analyses of large series of well-described patients.

Sommer distinguished postepileptic stupidity and episodic postepileptic insanity. Postepileptic stupidity was the final consequence of a dementing process which was initiated by recurrent postictal twilight states and consecutively followed by progressive forgetfulness, circumstantiality, word finding difficulties, impairment of perception and changes in character with religiosity, hypocrisy, irascibility and irritability. With respect to episodic insanity, in 63 patients Sommer distinguished psychoses which occur preictaly (n=5), postictally (n=47), or instead of seizures (epileptic equivalents, n=11). In Sommer's opinion these episodes were directly related to epileptic activity, in contrast to the rare cases of combinations of epilepsy with true madness (schizophrenia), melancholy or paralysis.

The term status was used «whenever a seizure persists for sufficient length of time (subsequently defined with at least 30 to 60 minutes) or is repeated frequently enough to produce a fixed or enduring epileptic condition.» This definition is enshrined into the World Health Organization dictionary of epilepsy by Gastaut [8] as well as the Handbook of clinical neurology by Roger, Lob and Tassinari [9] and Handbook of electroencephalography and clinical neurophysiology by Gastaut and Tassinari [10]. Today, a widely accepted operational definition of status epilepticus is that of a «condition in which epileptic activity persists for 30 minutes or more, causing a wide spectrum of clinical symptoms, and with a highly variable pathophysiological, anatomical and etiological basis». It is important to note that this definition implicates that status is not simply a rapid repetition of seizures (in fact the word «seizure» is no longer retained) and as such a repetitive (iterative) version of ordinary epilepsy, but a condition (or a group of conditions) in its own right with distinctive pathophysiological features.

Today it is estimated that there are between 65,000 and 150,000 cases of status epilepticus in the United States each year [11], and that approximately 25% are non-convulsive [12-14]. At least 10% of epileptic patients suffer a status epilepticus during the course of their disease, and 50% of status epilepticus appears in patients with no known history of epilepsy [15]. Status epilepticus is present in nearly all epileptic syndromes, even idiopathic ones, although it is more frequent in cryptogenic and symptomatic forms. Whereas tonic-clonic status epilepticus is the best known type and its diagnosis is simple, partial status epilepticus presents a diagnostic difficulty, and complex partial status epilepticus presents yet a remarkable challenge. Particularly difficult is the differential diagnosis of complex partial status epilepticus and absence status epilepticus, above all the form termed «late-onset de novo absence status epilepticus,» which presents as confusional syndrome in the elderly [15].

Janz [16] summarized two variants of Temporal Lobe Epilepsy with psychotic manifestations; (1) long-lasting sensory or somatosensory or «psychic» seizures, and (2) epileptic twilight states with productive-psychotic signs and symptoms. The first is a type of partial seizure and the second is a type of status epilepticus.

Purpose

This work is aiming at recording and confirming the concept of existence of:

-Prolonged complex partial seizures status epilepticus (for weeks rather than minutes or days)

-Psychotic behavior as the only presenting manifestations

-Complete and persistent response of the Psychotic behavior to anti-epileptic drugs, alone.

Subjects and methods

Subjects

The subjects of this study are 37 patients (17 males and 20 females) and their age range from 14 to 36 years. They were collected from El-Mataryia Teaching Hospital, General Organization for Teaching Hospitals and Institutes, Cairo, Egypt.

They present with psychotic manifestations such as:

- Formal thought disorder (37/37)
- Loss of insight, (37/37)
- Impaired judgment. (37/37)
- Delusions and hallucinations (29/37)
- Primary delusions (30/37)
- Excitement and aggressive behavior (25/37)
- Depersonalization and derealization (17/37)

They were suffering from the above manifestations for at least one week (up to four weeks), and drug naïve.

Methods: They were subjected to the following procedure:

- Thorough clinical examination was done after history taking.
- MMSE (Mini-Mental status examination) was done.
- EEG (ElectroEncephaloGram) was done first for diagnosis, and as needed later.
- Intravenous Bolus of diazepam (10 mg) was given.
- clinical reassessment, then,
- Anti-epileptic was prescribed, individually.
- Clinical assessment, in addition to MMS, was done every two weeks for the following three months, then monthly for the next twelve months.
- Brain imaging was done in some suspicious cases.

Results

Clinical examination was unremarkable; apart from subtle eye abnormal movement (e.g. increased frequency of blinking, nystagmus, stare look....etc).

MMSE score ranged from 26 to 13 at time of diagnosis. It returns to normal after 4 to 12 weeks of treatment

EEG of the patients showed:

- periodic lateralized left epileptiform activity in 13 patients (35%) figure (1);
- bilateral independent periodic lateralized epileptiform activity in 8 patients (22%) figure (2), and
- Generalized periodic epileptiform discharges in 16 patients (43%) figure (3).

IV diazepam improved 23 patients (62%) as indicated by improvement in the clinical picture, MMSE, and EEG picture.

Antiepileptic Drug Treatment:

•25 patients (67.6%) return to their normal after four weeks of monotherapy drug treatment.

•28 patients (75.7%) return to their normal after eight weeks of monotherapy drug treatment.

•The rest (9 patients, 24%) needed addition of another antiepileptic to be controlled.

Discussion

After a period of neglect, psychoses in epilepsy were 'rediscovered' in the 1950s and 1960s [17]. English and American authors as Gibbs, Gibbs and Fuster [18]; and Slater, Beard and Glithero [19] emphasized again the link to temporal lobe epilepsy (TLE).

Psychiatrists in the early twentieth century, like Sommer, stressed the similarity between psychosis in epilepsy and schizophrenia and therefore favored a positive relationship between both disorders. Since then the growing international interest in links between psychosis and epilepsy has generated a multitude of studies.

A classification system for psychosis in epilepsy should ideally consider psychopathology, duration and course of psychosis, type of epilepsy, relation to seizure or status activity, drug treatment, EEG findings and psychosocial factors. Unfortunately, these criteria are not strictly inter-correlated "Atypical" combinations are not unusual, such as ictal and postictal psychosis in clear consciousness [20]. Variations in phenomenology and precipitation can also be seen intra-individually in patients who experience recurrent psychotic episodes [20].

For the time being a comprehensive description considering each of the above-mentioned criteria separately is recommended. Epileptic syndromes and seizures should be classified according to the proposals of ILAE. Psychosis should also be classified following international systems such as the lCD 10 or DSM IV. However, when applied in psychosis in epilepsy, these classifications have some problems, which necessitate compromise. In the DSM IV, for example, the existence of epilepsy automatically leads to a diagnosis "organic mental syndromes and disorders" and does not allow the diagnosis of "functional" psychosis such as schizophrenia, even if all required criteria are met [21].

The psychotic syndromes seen can be classified according to their relation to seizures as such: Ictal [21], Postictal [21], Peri-ictal [22, 23], Interictal [21-23], and Alternative Psychoses [17,20,21].

Epileptologists have spent much effort in studying the phenomenological variety of seizures arising from the temporal lobe. The recent proposal for classification of the ILAE is more discouraging than helpful for clinical needs. It suggests that reliable classifications requires sophisticated, often invasive EEG methods because clinical seizure symptomatology is not specific and can therefore be misleading as can the routine scalp EEG. Limbic epilepsy, for example, which arises from a recognized functional system [23], has no place in the classification because it crosses anatomical boundaries. In many studies frontal and temporal lobe epilepsies are not sufficiently differentiated.

Complex hallucinations and delusions have been described as frequent aura symptoms of psychotic patients [23]. Kristensen and Sindrup [24] found an excess of dysmnesic and epigastric auras in their psychotic group and emphasized the localizing significance with respect to:

* Periamygdalar dysfunction.

*There was a significant relationship with the presence of both complex focal seizures and absence seizures.

This was interpreted as an indication of widespread dysfunction and as being of more significance than a localizable pathology since simple seizure symptomatology is more reliable in terms of localization than complex focal seizure symptomatology [25]. Kristensen and Sindrup [24] found a higher rate of ictal amnesia among their psychotic patients, which again might indicate that ictal impairment of consciousness plays a role in the development of psychoses.

Moreover, psychomotor status often evolves from or alternates with aura continua (what was called simple partial status epilepticus [26-28], so that many overlaps between aura continua and psychomotor status exist in literature and cases are finally categorized according to their full blown semiology (i.e., as psychomotor status, although for a certain period of time they would fulfill the criteria of simple partial status epilepticus). So, it is better categorized as non-convulsive status epilepticus.

Major reviews on status epilepticus were the Santa Monica, 1980 California Conference [29] and the Seventeenth Annual Merritt-Putnam in Boston, which was published as a supplement to Epilepsia [30]. In the same year, 5 position papers on nonconvulsive status epilepticus were published in the Journal of Clinical Neurophysiology [31].

It is difficult to deny the intriguing possibility that some abnormal mental states (in epilepsy) are due to prolonged seizure activity. Although there is undisputed evidence that prolonged epileptiform EEG discharges (characteristic of status) in hippocampal and amygdaloid regions can be associated with behavioral abnormalities and can occur with or without clear-cut scalp EEG changes, it is quite unknown to what extent the generality of «interictal behavioral peculiarities» might be associated with such «subclinical EEG status activity» in deep structures.

Since limbic status epilepticus implies seizure discharges in the limbic system, it is not surprising that without intracranial recording from the core structures of the limbic system such as hippocampal formation and amygdala, limbic status epilepticus is often undetectable. This might be one reason that non-convulsive status epilepticus is rarely reported in literature in comparison to convulsive

status epilepticus.
In Shorvon's suggested «revised classification of status epilepticus, we find the category «nonconvulsive» status. The term «electrographic» as a characteristic status seizure type is found in boundary syndromes «including electrographic status epilepticus with subtle clinical signs, prolonged postictal confusional status, the syndrome of acquired epileptic aphasia, and epileptic behavioral disturbances and psychosis [32].

Seven percent of epileptic patients, especially those with temporal lobe epilepsy, complain of psychiatric symptoms. They visited a psychiatry clinic, at least a year before diagnosis [23].
EEG changes were detected in 50-70% of acute and chronic schizophrenics, in many epidemiological studies [33].

Conclusion

In the present study a prolonged state of behavioral disturbance proved to be a special type of non-convulsive status epilepticus; lasts for at least a week; and, it was electrographically proven in accordance with the study results of Brenner [34] who described the most frequent EEG pictures in non-convulsive status epilepticus. Concluding that there is a special type of non-convulsive status epilepticus, which present with psychotic behavior, and, lasts for weeks. This may be a condition in its own right with distinctive pathophysiological features.

Depending on the diagnosis, the study patients' group received only antiepileptic drug treatment, which was tailored individually. Drug treatment proved to be effective. This type of status can be misdiagnosed as Brief psychotic disorder; if clinical examination (including MMSE) and EEG was not done for every suspicious case. Monotherapy is effective in 75.7% of cases. We can call this type of non-convulsive status A "Status Psychosus epilepticus".

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