

SHORT REPORT

Prolonged sensory or visceral symptoms: an under-diagnosed form of non-convulsive focal (simple partial) status epilepticus

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Abstract

Four patients had prolonged, sensory, simple partial seizures (SPS), lasting up to several days, without associated behavioural impairment. In three patients, the SPS often occurred as a prolonged "aura" before a more overt seizure. Descriptions included: "butterflies", rising epigastric sensation; "a thought in the stomach", and an olfactory sensation. Seizure localisation was frontal in one case, temporal in two cases and uncertain in one case. These sensations may represent an under-reported form of continuous, focal seizure activity, which arises from various cerebral regions.

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The epileptic aura is generally considered to be a short-lived phenomenon, representing the early stages of build up of abnormal, synchronous electrical activity. This usually develops further into a more obvious seizure or else halts abruptly, with reversion to the interictal state. We present four cases of prolonged, sensory SPS, without invariable ictal progression, presenting to us over the course of one year.

Case reports

Case 1: "Butterfly sensation"

This was a twenty three year old, right handed male motor mechanic. Complex partial seizures (CPS) first occurred when he was six years old, in association with fever and a head injury but they did not become regular until he was 10. They were frequently preceded, for up to two days, by a butterfly sensation, often felt in the abdomen but not specifically localised, which sometimes persisted after the seizure. Attacks were recorded with video-EEG-telemetry, before and after stereotactic EEG electrode insertion. They started with dizziness and pallor, followed by a single loud scream, abduction of his legs, bilateral upper limb posturing, then rocking movements and complex motor activity, for example, bicycling movements. After their recurrence, CPS occurred in clusters lasting up to one week, with a cluster often followed by a generalised tonic-clonic seizure (GTCS).

Ictal scalp EEG recording showed a 20 second build up of high amplitude sharp and

slow waves, negative in the right frontocentral leads. Postictally the continuation of the butterfly sensation was characterised by a period of frontocentral, electrographic status epilepticus (figure). During this episode there was no objective behavioural abnormality.

Intracranial ictal EEG revealed a build up of spike and wave at the deepest medial frontal electrode, which spread to the rest of the frontal lobe, to the temporal neocortex and to the hippocampus. There were frequent, widespread, interictal spikes that were not clearly correlated with the butterfly sensation. He did not, however, experience such a prolonged feeling as during scalp recording.

He had right frontal lobectomy with intra-operative electrocorticography. The resected tissue showed histological evidence of a neuronal maturation defect. One year later, he had suffered a single cluster of seizures, and no further prolonged auras.

Case 2: "Thought in the stomach"

Case 2, a twenty five year old, right handed male motor mechanic had a normal neonatal and early childhood history. At five years of age he suffered a GTCS without obvious precipitant. He was treated with phenobarbitone, and had no further attacks until the age of eleven years, when he started to experience CPS. These started with an aura of a "thought in his stomach", lasting up to one minute. A brief speech arrest with blankness, was followed by hand rubbing automatisms, dysphasia and amnesia, lasting a few minutes. He was seizure free from 16-20 years of age, but seizures then recurred in the same pattern. When he was 24 years old, his medication was changed and his overt seizures ceased. However, at roughly monthly intervals (the same frequency as his seizures one year previously) he experiences the same sensation of a "thought in his stomach". This fluctuates over several days, not disappearing and not reaching the severity he associates with an impending attack. During this time he remains fully alert and able to carry out his normal work and duties.

Interictal EEG consistently showed mild sharp and slow wave abnormalities over the right anterior temporal region. CT and MRI were normal. We have not obtained an EEG during an aura.

Case 3: Epigastric sensation

This was a 36 year old right handed, male housing officer with a normal perinatal and

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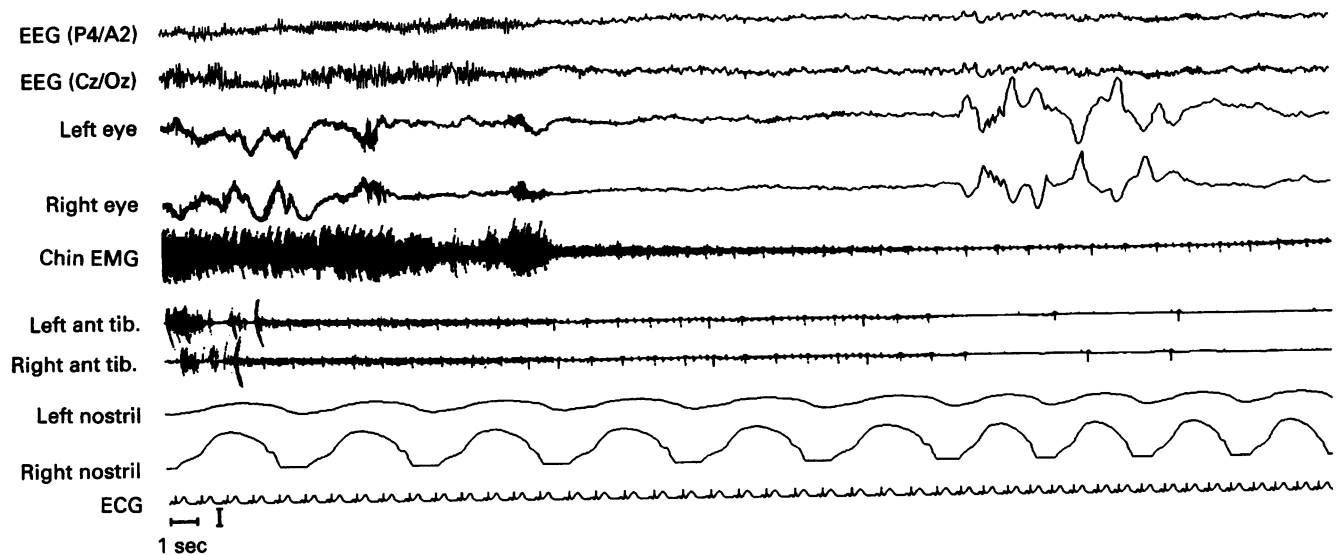


Figure 1 Case 1: Standard 10–20 scalp EEG during a prolonged epigastric sensation, showing rhythmical slow activity, predominantly in the right frontocentral leads.

early childhood history. Seizures started at the age of 18 and have been present once a week for the past 13 years. On the day he is due to have a seizure, he awakes with an epigastric sensation, which persists for hours and a few seconds before an attack, it rapidly rises to his chest. He shouts “Oh God” and is unaware for several seconds. There follows up to one minute of manual automatisms. Recovery occurs within two minutes, with no dysphasia. Sometimes the epigastric sensation persists after the seizure when he knows he will have a further seizure that day.

CT scan was normal but MRI, with planes orientated parallel to the long hippocampal axis, showed substantial loss of right hippocampal substance. Interictal EEG on a normal day showed diffuse delta and sharper components anteriorly with a left-sided preponderance. Video telemetry was performed: there was no EEG abnormality during an aura or a seizure, but postictally there was diffuse right sided slow activity.

Case 4: Olfactory sensation

A right handed 55 year old male electronics engineer had experienced occasional micropsia, with the illusion of progressive diminution of the size of objects since the age of four years. This lasted for two minutes, with no alteration of consciousness and was associated with a pleasurable sensation. Since 1984 he has suffered frequent, strong, olfactory hallucinations of a variable nature, but usually a chemical odour, resembling paint or hot oil. These would persist for several days with numerous exacerbations lasting up to 30 minutes. There have not been any more overt seizures and his work and concentration remain unimpaired.

Interictal EEG consistently showed a left-sided spike focus between the anterior and midtemporal regions, which did not change in association with the hallucinations. CT scan was normal but volumetric MRI showed reduction in volume of the anterior portion of the left hippocampus. Initial treatment with

carbamazepine made no impact on his attacks, and subsequently phenytoin was only transiently effective.

Discussion

An aura represents a focal epileptic discharge, manifesting as a sensory SPS, which may progress to a more overt seizure. The first three cases clearly satisfy the International League Against Epilepsy criteria for an aura;¹ a stereotyped sensation, without impairment of consciousness, temporally closely related to seizures. The fourth is also epileptic in view of clinical features, EEG and MRI findings and response to anticonvulsants. They are, however, unusual in that the phenomena may persist for a considerable period without development of an overt seizure. This can be classified as “simple partial status epilepticus”, non-convulsive as distinct from *epilepsia partialis continua* (EPC) or other motor seizure phenomena.

For case 1, the sensation was clearly associated with electrical seizure activity on surface but not on intracranial EEG. Up to half of auras may be missed on intracranial recording, probably because of the small volume of tissue sampled.^{2,3} The diagnosis of an aura is clinical; not affected by the absence of localising EEG abnormality, either ictal (cases 3, 4) or interictal (case 2).^{3,4}

Evidence for a frontal focus is strong in case 1, and cases 3 and 4 strongly suggest temporal lobe epilepsy. These results present evidence that prolonged, sensory SPS is not a property of a single cerebral region.

Psychomotor complex partial and focal motor status epilepticus are well accepted entities, but despite series amounting to thousands of cases with careful analysis of seizure semiology,^{3,5,9} we have found few reports of sensory status epilepticus. Hughlings-Jackson¹⁰ described a case in which: “sometimes when the attacks are about, she has a sensation of smell or of taste at the back of her throat, but

not in connection with her seizures". Penfield and Jasper¹¹ described a patient with a previous parietal injury who suffered continual tingling of the left foot for a week and recently, Sowa and Pituck¹² reported a case with weeks of continuous visual hallucination, accompanied by EEG abnormality and with no functional impairment. Our four patients were taken from our current clinical practice and we have seen others with a past history of a similar phenomenon, suggesting that this pattern of epilepsy may be more frequent than previously recognised. These patients with a prolonged, continuous SPS are distinct from others who experience frequent, brief auras before an overt seizure.

These symptoms represent continuous, highly localised seizure activity, manifesting as sensory or visceral symptoms, without spread to areas that would result in behavioural impairment. The subtlety of these symptoms, that they last longer than is usual for epileptic phenomena and that they are rarely accompanied by EEG change, probably explain why they are less frequently recognised than other forms of focal status epilepticus, for example, EPC or psychomotor. This may mean that a group of patients has undetected, mildly symptomatic, prolonged seizure activity; case 4 had symptoms from early childhood, but a diagnosis was only made in middle age.

Apart from patient 1, who also had severe complex partial seizures, all these patients were in employment and continuing to function normally in their social setting. This may

therefore be a relatively benign form of prolonged seizure activity. If, as has been suggested, continuous, focal discharges are harmful to neurons in the long term,¹³ then early identification and treatment of this group may prevent the development of a more intractable seizure disorder, and associated neuropsychological deterioration.

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patients with chronic lumbosacral disc diseases as described in our study. We think there are important differences in pathophysiology. So far the testing of neuropathic conditions with heat pain stimulation has rarely resulted in strong evidence for hyperalgesic or hyperpathic changes. In a very recent publication,⁷ however, Wall gave a great number of examples of hyperalgesic changes produced by different kinds of neuropathies and he also pointed to the fact that a non-selective blockade of peripheral afferent impulses may lead to a "partial disinhibition" and, in consequence, to hyperalgesia. This is what seemed to have happened in our patients with chronic lumbosacral disc disease. That such an event might produce effects at the contralateral side appears not too speculative when the results of contralateral TENS-effects cited in our paper are considered. Taken together, we still believe that the conclusions drawn from the pilot study described are justified.

Finally, we want to answer the questions raised by Bowsher. Two patients were affected at the L5 root affection and 7 at the S1 root. We measured the thresholds at the medial (L5) and lateral (S1) side of the dorsum pedis, where the peripheral dermatomes are to be found, and verified the location of the dermatomes in the preceding neurological examination. As Bowsher expected, the dermatomes do normally not differ in warmth and pain sensitivity.

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NOTICES

Tropical Neurology Symposium, London, 25 March 1993

The first Tropical Neurology symposium to take place in the United Kingdom will be held at Manson House, 26 Portland Place, London. It will be co-sponsored by the Tropical Neurology Research Group of the World Federation of Neurology jointly with the Royal Society of Tropical Medicine and Hygiene. It will be chaired by Lord Walton, Professor D Warrell, Professor N Wadia and Dr CM Poser.

Further details from: The Administrator, Royal Society of Tropical Medicine and Hygiene, Manson House, 26 Portland Place, London W1N 4EY. Telephone: 071-580 2127; Fax: 071-436 1389.

First Congress of the International Stereotactic Radiosurgery Society, Stockholm, Sweden

This congress will be held from 16-19 June 1993. Further information from The Con-

gress Secretariat ISRS 1993, Stockholm Convention Bureau, PO Box 6911, S-102 38 Stockholm, Sweden.

Ciba-Geigy · ILAE · IBE · Epileptology Prize

Ciba-Geigy has agreed with the International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE) to establish a new prize of SFr. 20.000. to be awarded in recognition of outstanding achievement in the field of epilepsy. The prize is designed to encourage applied human research in epilepsy. By cooperating with both ILAE and IBE, Ciba-Geigy emphasises that application for the prize is open to candidates from all fields of applied research.

Anyone outside the pharmaceutical industry who considers that he or she has made a significant scientific contribution in the field of epilepsy may compete for the prize. Deadline for submission is 15 January 1993.

Entries for the prize will be judged by an Adjudicatory Panel consisting of the Presidents and one further delegate each of ILAE and IBE, and an independent chairman.

The prize will be awarded at the opening ceremony of the International Epilepsy Congresses (for the first time in Oslo on 3 July 1993, and subsequently in Sydney in 1995).

For further details and a set of the rules and application forms, those interested should write to Mrs. G Haldemann, Ciba-Geigy Limited, CH-4002 Basel, Switzerland.

Correction

In the article by M Manford and SD Shorvon, Prolonged sensory or visceral symp-

oms: an under-diagnosed form of non-convulsive focal (simple partial) status epilepticus (August 1992 issue of the jour-

nal), the figure was incorrect. The figure should have been as follows:

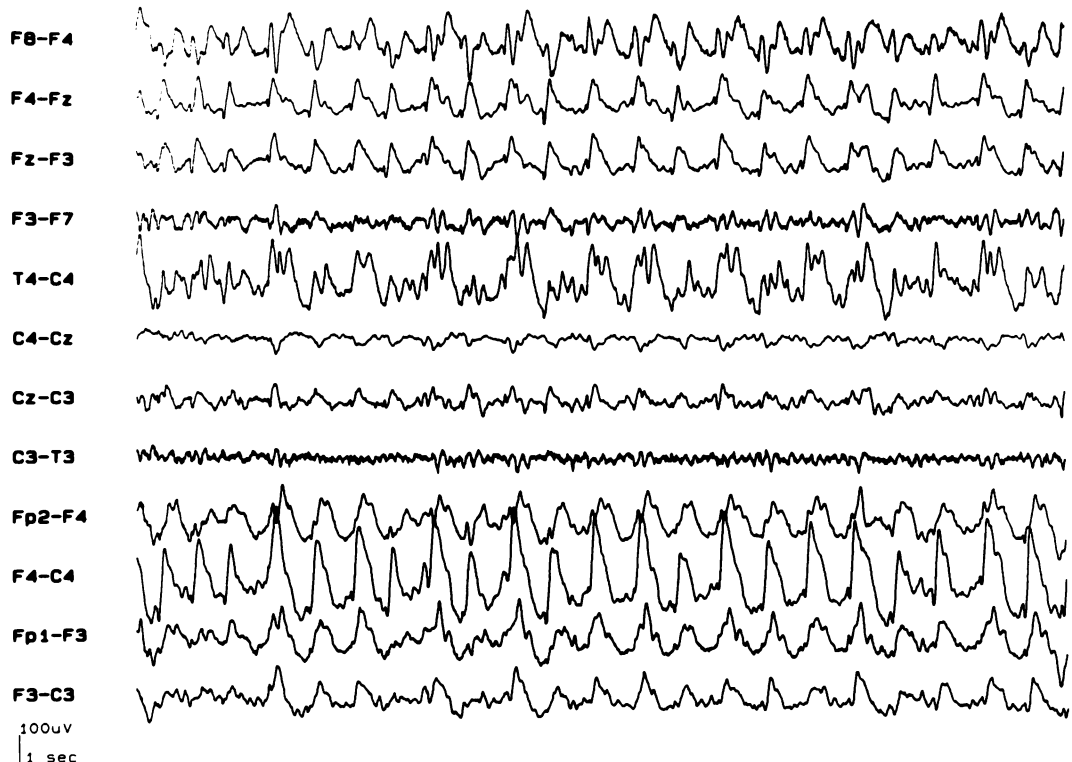


Figure Case 1: Standard 10-20 scalp EEG during a prolonged epigastric sensation, showing rhythmical slow activity, predominantly in the right frontocentral leads.