The syndrome of isolated epileptic status¹

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SUMMARY Within a group of 23 patients who presented with status epilepticus a syndrome is defined in which sudden unheralded status occurs in apparently healthy individuals. Recovery from the attack is complete and no other evidence of cerebral pathology may be found at the time. In seven of nine such cases studied the final diagnosis at follow-up, either by necropsy or by operation, was of cerebral tumour. In five of the seven the fronto-temporal region was the site of pathology. It is suggested that the occurrence of isolated status indicates a possible cerebral tumour for which careful search should be made and, if negative, follow-up study arranged. Of 20 of our patients with status epilepticus in whom the site of lesion was definite, nine were exclusively frontal and a further six had some frontal involvement. This confirms previous evidence that symptomatic status epilepticus indicates a frontal lesion.

From the records of 2,500 patients with seizures seen in United Oxford Hospitals between 1947 and 1969, we obtained 166 cases of status epilepticus for analysis.

Our definition of status was either that three or more grand mal convulsions occurred without intervening recovery of consciousness, or that there was at least an hour of continuous convulsions without recovery of consciousness. These criteria were rigidly applied, so that we probably excluded some genuine cases of status where the evidence was uncertain.

Of the 166 cases accepted, for reasons concerning investigation and management, we considered further only 96 patients aged 11 years or over. The age incidence is shown in Table 1. Table 2 shows the probable causes for the condition. The incidence of symptomatic status—about two-thirds of the cases—is relatively high, as is the case in most other series. The high mortality in these symptomatic cases is also noteworthy: including deaths as an immediate result of status, some 50% of the cases died within six months.

We wish to discuss 23 of the cases who presented with status as their initial symptom. They are divided into two groups. In the first group of 14 cases there was either an already known pathology, or this was established at the time of the occurrence of status. All these cases are therefore examples of symptomatic

TABLE 1
AGE AT FIRST STATUS EPILEPTICUS

<i>Years</i>	<i>No. of patients</i>
11-20	19
21-30	21
31-40	15
41-50	6
51-60	16
61-70	15
71-80	4
Total	96

TABLE 2
AETIOLOGY OF STATUS EPILEPTICUS IN 96 CASES
AGED 11 TO 80 YEARS

<i>Pathology</i>	<i>No. of patients</i>
Unknown and idiopathic	32
Trauma	14
Tumours	19
Degenerative and arteriosclerotic	13
Infectious	9
Congenital and infantile brain lesion	4
Miscellaneous	5
Total	96

status. Table 3 shows the pathological lesion in these cases. In all but one of them it was primarily cerebral.

There is a further group of nine cases for which we suggest the term 'lone status epilepticus.' The

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TABLE 3
EPILEPSY PRESENTING AS STATUS OF KNOWN CAUSE

Pathology	No. of patients
Degenerative	3
Brain scar	3
Encephalitis	2
Carcinomatosis	2
Angioma	1
Hyponatraemia	1
Syphilis	1
Anoxic	1
Total	14

main features of this syndrome are: (1) the status presents abruptly in an otherwise healthy adult, (2) there is no past history of any neurological illness, (3) there is complete recovery from the attack, (4) investigations at the time of the attack reveal no causative lesion.

Table 4 shows the age at onset and the investigations carried out. In only one case was there a localized EEG abnormality and, in this case, lumbar puncture and pneumoencephalograms were normal. In two cases the protein was raised and in one of them (case 5) further investigation, which was refused, might have revealed the pathology discovered some six months later. In the other case (case 9), a symmetrical bilateral EEG abnormality led us to postpone further investigation. The patient remained well for 18 months but was then lost to follow-up.

Table 5 shows the subsequent course of the cases and the final diagnosis, achieved either at necropsy or at operation. Further occasional isolated grand mal attacks occurred in three cases, but the emergence of signs of local cerebral disease led to the diagnosis in the majority. Seven of them had proven

TABLE 4
LONE STATUS EPILEPTICUS: INVESTIGATIONS AT TIME OF ATTACK

Case no.	Age at onset (yr)	EEG	CSF	Angio or AEG
1	30	Moderate dysrhythmia	Normal	—
2	29	Moderate dysrhythmia	Normal	Normal
3	28	Non-focal slow waves	Normal	Normal
4	65	Mild dysrhythmia	Normal	—
5	50	—	Protein 100 mg	—
6	53	R. frontal spike focus	Normal	Normal
7	59	Moderate dysrhythmia	—	—
8	73	Mild dysrhythmia	Normal	—
9	50	Bilateral synchronous 3 Hz	Protein 80 mg	—

TABLE 5
LONE EPILEPTIC STATUS: AETIOLOGY AND INTERVAL BEFORE DIAGNOSIS

Case no.	Final diagnosis	Interval before diagnosis (yr)	Method of diagnosis
1	L. frontal astrocytoma	9	Operation
2	R. frontal astrocytoma	5	Operation
3	L. parietal oligodendroglioma	5	Operation
4	Glioblastoma	11	Necropsy
5	R. parieto-temporal astrocytoma	6/12	Operation
6	R. frontal glioblastoma	4/12	Operation and necropsy
7	L. temporal glioblastoma	3/12	Operation
8	Cerebral atrophy	9	Necropsy
9	Untraced	?	?

cerebral tumours. Five of these involved frontal and/or temporal regions. In one, the site was indefinite but described at necropsy as in the anterior part of the hemisphere. The interval from initial status to a verified diagnosis varied from three months to nine years.

Thus we would add a fifth feature to the syndrome: cerebral tumour as its likely cause. The tumours were of varying grades of malignancy; from an oligodendroglioma diagnosed nine years later to a glioblastoma declaring itself in three months. They tended to be anteriorly situated in the hemispheres but were not exclusively so.

Although there are isolated references to sudden status epilepticus as a presenting feature of frontal tumours, we have found no previous description of any group of cases with the picture we have given. In investigating cases of status occurring in non-penetrating brain injuries, one of us found evidence of frontal lesions in 15 of 17 cases. Janz (1964) also noted that, in a series of cerebral tumours in 27 of which status occurred as part of the symptomatology, 20 or 74% were frontally situated. Further, in 97 cases of epilepsy from penetrating brain injuries, he also showed that 94% of those which had status epilepticus had frontal injuries. In this excellent study devoted to status epilepticus and frontal lobe lesions, a number of the cases mentioned would fall into the category we are describing. One of us earlier noted also (Whitty, 1960) that in a series of 356 gunshot wounds of the brain, showing post-traumatic epilepsy, 14 had status as their only form of epilepsy. Twelve of these were frontal wounds.

Pursuing this point in our present status cases, there were 20 patients of the total 96 in whom the site of lesion was definite. The findings are shown in Table 6. Fifty per cent of them were exclusively

TABLE 6
SITE OF LESION IN 20 CASES OF STATUS EPILEPTICUS

Site	No. of patients
Frontal	9
Fronto-temporal	4
Fronto-parieto-temporal	2
Temporal	2
Parietal	3
Total	20

frontal, and 75% involved the frontal lobes as well as other areas.

We therefore conclude that:

1. Confirming earlier findings, symptomatic status

indicates a frontal lesion. This is both of diagnostic value and of anatomical interest as implying elective pathways of spread of discharge from the frontal lobes.

2. The occurrence of lone status epilepticus indicates the probability of a cerebral tumour and should guide further diagnostic search and influence prognosis.

3. Status epilepticus—especially symptomatic—is still a serious condition with a high mortality.

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