Letters

This is the first case of progressive supranuclear palsy in which falls, which is a characteristic early symptom of the disease, are clearly due to focal paroxysmal kinesigenic choreoathetosis and they have many of the criteria proposed by Kertesz. These criteria are: duration and character of the paroxysms, response to anticonvulsants even though the dystonic illness was far advanced and more complex in this case, the precipitating factor and absence of EEG abnormalities. The late age of onset of this focal paroxysmal kinesigenic choreoathetosis and the absence of a positive family history is explained by the “symptomatic” nature of this disorder. However, later in the evolution of the tonic disorder in this case of progressive supranuclear palsy, the dystonia became more or less permanent. This may explain the inability of levodopa alone to abolish this more complicated dystonia as in the case described by Loong although it effectively resolved the persistent plastic rigidity component. At this point the paroxysmal component of this disorder became apparent again as in earlier stages and addition of carbamazepine effectively abolished the focal paroxysmal kinesigenic choreoathetosis. This is also a first report of progressive supranuclear palsy in Eastern Africa, which partly explains the delay in the diagnosis of this patient.

The occurrence of paroxysmal kinesigenic choreoathetosis in progressive supranuclear palsy is not surprising as most recent authors of the former consider it to be a disorder of basal ganglia function despite the only necropsied case showed no definite pathological findings, while in the later, pathological lesions in the basal ganglia are prolific. Finally, it is recommended that in patients with Steele-Richardson-Olszewski syndrome who fall, or in any obscure falls, focal paroxysmal kinesigenic choreoathetosis should be seriously considered and if this disorder is suspected a trial of carbamazepine should be instituted.

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References


Status epileptics due to abrupt diazepam withdrawal: a case report

Sir: Benzodiazepines are extensively used as anxiolytic and hypnotic drugs and are usually considered as having a low risk for pharmacological dependence. However, many authors have warned of the existence of a benzodiazepine withdrawal syndrome. We report a patient, heavily dependent on diazepam, who had a confusional psychosis and generalised tonic-clonic status epileptics a few days after the abrupt discontinuation of the drug.

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Fig EEG recorded during the episode of generalised convulsive status epileptics. Recruiting rhythms at 11–13 Hz (a) followed by generalised spike- and polyspike-wave discharges ending with a short phase of bioelectric depression (b). During the seizure only slight motor manifestations of convulsive activity were observed (for detail see text).
extraventricular. During the intervals the patient was in a deep comatose state. The EEG showed, besides a diffuse slowing of the background activity, bursts of generalised 11–13 Hz recruiting rhythms of short duration, followed by generalised spike- and polyspike-wave discharges ending with short phases of bioelectric depression (fig). A CT scan and a lumbar puncture gave normal results. Intravenous phenytoin (750 mg/day) and phenobarbital (200 mg/day) were started. The patient gradually improved, even though less frequent seizures were still observed over the next 24 hours. On the third day he was fully alert and was transferred to our Centre for Epilepsy. EEG abnormalities (short generalised discharges of irregular spike-wave complexes and a moderate diffuse slowing of background activity) gradually improved. When the patient was discharged, on the 30th day, he was taking phenytoin (300 mg/day) and mephenobarbital (120 mg/day) and, over the next three years, he has been in good health.

The patient and his relatives gave no personal or family history of epilepsy or seizures or of alcohol abuse. However, they revealed that the patient, who was a medical student, for many years suffered from anxiou nervousness and depression, partly due to his failure in school. For this reason he used to take very high dosages of benzodiazepines. During the 7 months which preceded the convulsive status epilepticus he habitually took 40–50 tablets of 5 mg diazepam (200–250 mg) every day. Since symptoms of anxiety did not cease and drowsiness and disturbances of the gait had appeared, the patient was examined by a psychiatrist who prescribed an antidepressant (maprotiline chlorhydrate, 50 mg) and recommended discontinuing diazepam, which the patient did abruptly. During the following 4 days he was especially agitated and anxious, managed to sleep only a few hours a night, experiencing terrifying dreams which, particularly during the last two days, he could not distinguish well from reality. Occasionally he had myoclonic jerks of the upper extremities, was confused and felt persecuted. On the fourth day after diazepam withdrawal, in the morning, about an hour after awakening, he had his first generalised tonic-clonic seizure.

It is known that treatment with benzodiazepines over prolonged periods can lead to physical dependence and a characteristic withdrawal syndrome.2–4 Seizures,5–8 confusional or paranoid psychoses9 and coma10,11 are the most severe withdrawal symptoms reported and are more frequently observed 2–10 days after abrupt discon-continuation of high dosages and/or long term treatment.4

In our patient the most commonly observed diazepam withdrawal symptoms (insomnia, increasing anxiety and restlessness) preceded a confusional state and then the convulsive status epilepticus. The temporal relationship and the clinical picture firmly supported the hypothesis that the patient’s symptomatology had been caused by the abrupt diazepam withdrawal. In fact, the administration of 10 mg of diazepam IV, when our patient was confused, produced an almost complete, although transient, disappearance of his symptoms. However, repeated administration of the drug was subsequently unable to control seizures and to prevent the occurrence of status epilepticus. We have not a clear explanation for this phenomenon although tolerance may be involved. The administration of maprotiline after diazepam withdrawal might have been a factor in causing the acute disorder of our patient. Antidepressants can lower the threshold for seizures and in some cases were considered to be a contributing factor of benzodiazepine-withdrawal symptoms.11,12

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References


Recurrent neuroleptic malignant syndrome and hyponatraemia

Sir: The neuroleptic malignant syndrome is an uncommon acute or subacute reaction to neuroleptic medication characterised by pyrexia, muscle rigidity, altered consciousness and elevated creatine kinase levels.1 It is also cause is unknown, additional triggers and prevent not described and recurrence has not been reported, despite reintroduction of neurolepsy therapy on frequent occasions after single episodes. We describe a patient with recurrent neuroleptic malignant syndrome associated with hyponatraemia.

In 1958 a 20-year-old man developed paranoid schizophrenia. Over the next years he received electroconvulsive therapy (ECT), chlorpromazine and lithium, haloperidol, trifluoperazine and fluphenazine decanoate. Excessive water drinking was documented on two occasions. At the age of 43 years he was prescribed only fluphenixol decanoate 80 mg fortnightly, trifluoperazine 20 mg and orphenadrine 100 mg daily.

In February 1983, aged 44 years, he had three generalised seizures. Four hours later his axillary temperature was 38.2°C and severe generalised rigidity was noted. The serum sodium (Na+) was 112 mmol/l, potassium (K+) 1.9 mmol/l, osmolality 269 mOsm/kg, and urine osmolality 450 mOsm/kg. The white blood cell (WBC) count rose to 15.6 × 10⁸/l and aspartate transaminase (AST) to 450 IU/l. Cerebrospinal fluid (CSF) examination was normal. Intravenous potassium infusions were given and fluids restricted. Ten days later serum Na+ was 134 mmol/l and K+ 2.9 mmol/l. Previous medication was restarted.


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