she exhibited various automatism. However, as her illness progressed this variability became less obvious and any possible cyclicity still present was on a day to day level rather than hour to hour.

The EEG recordings have always shown bilateral slow wave activity with multi-focal sharp waves and spikes but no apparent rhythmic patterns or paroxysms. This picture is consistent with CPSE of longer duration. In the earlier stages and at seizure onset the picture is usually one of recurring cycles of activity similar to that seen in isolated complex partial seizures.

It has recently been demonstrated that CPSE can originate in the frontal lobes and, in fact, many previous reports of CPSE lend support to this when reviewed retrospectively.¹² We were unable to record the onset of any of our patient's five episodes of status. However, on reviewing the clinical features of the seizure onsets, it is possible that the 2nd, 4th and 5th episodes may have had a frontal origin.

There is increasing experimental evidence that repeated electrical discharges may produce neuronal damage, even in the absence of motor convulsive activity.^{13,14} In our patient a biopsy of the frontal region after 5 months of CPSE merely showed non-specific changes. There had been a 48 hour period of recurrent episodes of grand mal status earlier in the illness. The changes were not necessarily directly due to the CPSE.

Of the approximately 60 cases of CPSE documented in the literature so far, five have had impairment of higher mental functioning persisting after the attack.^{6,8,9,15} Three of these^{6,15} had as their main feature a marked impairment of memory function. One was the atypical case of CPSE mentioned earlier.⁹ The area of deficit in this case was not specified. One was a 60 year old diabetic male with "aphasic status epilepticus", in whom the authors believed there had probably been a small cerebral infarct during the ictal activity.⁸ In our patient neuro-psychological testing demonstrated that memory was severely impaired in the first few weeks after the episode had terminated, but that memory function is now probably approaching the pre-morbid level. There are still mild deficits in verbal fluency, visual memory and constructional skills, and the patient remains mildly disinhibited, possibly reflecting the frontal lobe damage.

The increasing reports of persisting post-ictal impairment has further consequences. The importance of accurate case definition is paramount and in their recent review Treiman and Delgado-Escueta¹¹ suggested

that a patient can be considered to be in CPSE if there are repeated complex partial seizures without full recovery between seizures to a completely normal state with no signs of residual deficit. This statement, therefore, must be revised since full recovery between attacks can no longer be a requisite to stating that the attack has terminated.

Myoclonus was a prominent feature of the illness in our patient and reflex myoclonus also occurred in the latter stages. Action myoclonus may occur after cerebral anoxia, but we feel that this was unlikely to be the cause in our patient since the myoclonus eventually subsided. Movement disorders may also occur with anticonvulsant usage; however, our anticonvulsant levels were never in the toxic range, although we were not able to measure free levels. It is not possible to be sure whether the myoclonus was due to the anticonvulsants or the CPSE. However, we favour the latter explanation.

There is no rigid strategy of drug therapy for terminating CPSE except that diazepam is the drug which most often successfully ends attacks. The recommended treatment at present is intravenous diazepam and loading with phenytoin. On review of other cases of CPSE there does not appear to be any other drug which is regularly successful in terminating episodes of CPSE. It seems that our patient's attack terminated when barbiturates were introduced.

In conclusion, our case serves to reinforce further the point that CPSE should be treated vigorously at the earliest possible opportunity, since it may convert to major motor status and even in the absence of this CPSE on its own may produce persisting changes in higher mental functions after the attack has terminated. The illness in our patient was very prolonged and latterly we felt that we must have been dealing with an irreversible and progressive process. On many occasions we felt that any further therapeutic manipulations would be futile. However, this was not so and the patient is now almost completely recovered.

MA ROBERTS

PRD HUMPHREY

*The Mersey Regional Department of
Medical and Surgical Neurology,
Walton Hospital,
Rice Lane,
Liverpool L9 1AE, UK.*

References

- De Pasquet E, Guadin E, Bianchi A, De Mendilaharsu S. Prolonged and monosymptomatic dysphasic status epilepticus. *Neurology* 1976;26:244-7.

- Hamanka T. Zum Problem des Status Psychomotorisches AnFalle. *Clin Psychiatry* 1972;14:592-603.
- Escueta AV, Boxley J, Stubbs N, Waddell G, Wilson WA. Prolonged twilight state and automatism: a case report. *Neurology* 1974;24:331-9.
- Hamilton NG, Mathews T. Aphasia: the sole manifestation of focal status epilepticus. *Neurology* 1979;29:745-8.
- Markand N, Wheeler GL, Pollack SL. Complex Partial Status Epilepticus (Psychomotor Status). *Neurology* 1978;28:189-96.
- Engel J, Ludwig BI, Fetell M. Prolonged CPSE: EEG and behavioural observations. *Neurology* 1978;28:863-9.
- Murasaki M. Psychomotor Status: case reports and proposal for classification. *Folia Psychiatr Neurol Jpn* 1979;33:353-7.
- Dinner D, Leuders H, Lederman R, Grettner T. Aphasic status epilepticus: a case report. *Neurology* 1981;31:888-90.
- Mikati MA, Lee WL, Robert De Long G. Protracted Epileptiform Encephalopathy: An unusual form of partial complex status epilepticus. *Epilepsia* 1985;26:563-71.
- Ballenger CE, King DW, Gallagher BB. Partial Complex Status Epilepticus. *Neurology* 1983;33:1545-52.
- Treiman DM, Delgado-Escueta AV. Complex Partial Status Epilepticus. In: Delgado-Escueta AV, Waskerlein CG, Treiman DM, Porter RJ, eds. *Advances in Neurology, Vol. 34: Status Epilepticus*. New York: Raven Press, 1983:69-81.
- Williamson PD, Spencer DD, Spencer SS, Novelly RA, Mattson RH. Complex partial status epilepticus: a depth electrode study. *Ann Neurol* 1985;18:647-54.
- Scheibel ME, Scheibel AB. Hippocampal pathology in temporal lobe epilepsy: a Golgi survey. In: Brazier M, ed. *Epilepsy, its Phenomena in Man*. New York: Academic Press, 1973:311-37.
- Collins RC, Lothman EW, Olney JW. Status epilepticus in the limbic system: Biochemical and Pathological changes. In: Delgado-Escueta AV, Waskerlein CG, Treiman DM, Porter RJ, eds. *Advances in Neurology, Vol. 34: Status Epilepticus*. New York: Raven Press, 1983:277-88.
- Treiman DM, Delgado-Escueta AV, Clark MA. Impairment of memory following prolonged CPSE. *Neurology* 1981;31:109 (abstract).
- Dunne JW, Summers QA, Stewart-Wynne EG. Non-convulsive status epilepticus: A prospective study in an Adult General Hospital. *Q J Med. New series* 62, 1987;238:117-26.

Accepted 6 July 1987

Akinetic mutism in Wernicke-Korsakoff disease: a case report

Sir: Akinetic mutism, first described by Cairns *et al.*,¹ may be seen in three types of

Letters

lesion,⁴ "all largely interfering with reticular-cortical integration but largely sparing corticospinal pathways: subacute communicating hydrocephalus; large bilateral, basal-medial frontal lobe lesions...; and tiny (and probably incomplete) lesions interrupting the reticular formation of the posterior diencephalon and adjacent mid-brain." Wernicke's encephalopathy⁷ has, to our knowledge, not yet been described as a causative factor in this condition; hence we report the following case.

A 74 year old woman was admitted to hospital on 5 August 1985 with dehydration, vertigo and cardiac arrhythmia. According to her family she was a heavy drinker. Five days before admission she had been walking around in her flat, talking with her daughter, who had come to see her. On the day of admission she was found lying in bed, motionless, open-eyed, whispering only "yes" when asked questions, and had been incontinent of urine and faeces. Her general physical condition was poor; body temperature was 35.9°C, arterial blood pressure was 140/80 mm Hg, she was not unconscious but did not answer questions. Hypokalaemia (2.8 mmol/l) and a minor metabolic acidosis were successfully treated by bicarbonate and a 5% glucose drip with potassium load. The next day her body temperature was 33.8°C; the bladder was catheterised for urinary incontinence. On the third day the patient was transferred to the neurological department. She lay motionless in bed, with eyes wide open, giving a distinct impression of wakefulness. She did not say a word, but after persistent questioning she gave her first name and her age (wrong by 30 years). No movement at all was executed on demand, vigorous nociceptive stimulation was followed by a slight and slow withdrawal of the limb concerned with neither vocal expression nor sign of emotion. There was very little facial reaction to strong pressure on the retromandibular branch of the facial nerve and no defence reaction at all to this manoeuvre. Body temperature was 32.4°C, without shivering. Oculomotor paralysis was almost complete, without voluntary movements, pursuit and doll's head phenomenon. Only minimal saccadic lateral movements of a few degrees persisted on pursuit; a moderate miosis without anisocoria was noted, both pupils reacted sluggishly to bright light. Moderate axial rigidity, and complete absence of all deep tendon reflexes was noted; plantar response was absent on the left, extensor on the right. Elevation of the uvula was absent, there was no deviation of the soft palate; drooling from inability to swallow saliva was present.

Arterial blood pressure was 100/50 mm Hg, and the pulse rate 80/min.

She was given 50 mg of thiamine intravenously once and 500 mg per day intramuscularly. Six hours later the akinetic and mute state had totally disappeared, body temperature was 37°C, but ocular movements took several days to recover, at first vertical then lateral movements. A severe memory impairment of Korsakoff type was diagnosed.

Six days after the start of thiamine therapy the patient became drowsy, answering slowly. Thiamine treatment had been stopped 3 days before by mistake and she had been on only 10 mg of thiamine in a polyvitamin preparation given with a glucose drip. Consciousness quickly returned to normal with proper thiamine treatment. Upon discharge six weeks later and at follow-up 15 months later ocular movements were full, consciousness normal, no bulbar signs or pathological brain stem reflexes were found, all deep tendon reflexes were present and symmetrical, plantar responses absent, position sense of the toe was slightly impaired, and there were no signs of a cerebellar syndrome. She still showed a Korsakoff syndrome and signs of urinary but not of faecal incontinence.

Blood transketolase activity was 14 milli-units/ml (normal: 60 ± 15 mu/ml) before thiamine treatment and 45 mu/ml after several days of treatment. The following results were normal: thyroid hormones T3, T4 and TSH, repeated blood cultures, blood glucose level, hepatic enzymes, creatinine, urea, blood electrophoresis and complete blood cell count (except for elevated MCV of 107 fl). Initial ECG: sinus rhythm, 100/min, ventricular extrasystoles, no J wave. A radionuclide brain scan with technetium 99m was normal, as was a high resolution, contrast enhanced CT scan, apart from diffuse brain atrophy (fig).

The diagnosis of Wernicke's encephalopathy in our case is proved by a history of chronic alcoholism, oculomotor paralysis, hypothermia, low blood transketolase activity, worsening under nutrition by glucose drip during hospitalisation and dramatic improvement by parenteral thiamine treatment. Subsequent memory impairment was of Korsakoff type. The impairment of consciousness was characterised by a distinctive impression of wakefulness, no spontaneous verbal or motor expression at all and only minimal reaction to noxious stimuli without any emotional participation. Intense solicitation resulted in rudimentary speech, testifying to intact motor speech function. Incontinence was complete, urinary and

Fig Contrast-enhanced CT scan: normal, apart from diffuse brain atrophy

faecal. It is important to note that this disorder of consciousness developed early in the course of the disease, no preceding coma had been noted and thus our case corresponds to the syndrome of akinetic mutism according to the criteria established by Plum and Posner.⁴ Normal consciousness returned promptly after thiamine treatment, thus strongly suggesting a causative relationship between the Wernicke's encephalopathy and the akinetic mutism. The disorder was clearly distinct from the "global confusional state" seen in a majority of cases with Wernicke's encephalopathy⁵ and from the comatose state in patients with hypothermia² insofar as those patients do not show the dissociation between an important hyporeactivity and an impression of wakefulness, typical of akinetic mutism. The association of Wernicke's encephalopathy and akinetic mutism has been suggested by Nielsen,³ though his description lacks detail. His patient, a 70 year old male alcoholic, was "conscious but unresponsive, attention to sensory stimuli was almost nil, on the third day of hospitalisation he responded with monosyllables, and when food was placed in his mouth he ate." A complete ophthalmoplegia and a bilateral Babinski sign were found. The patient died on the fourth day of hospitalisation of bronchopneumonia. At necropsy haemorrhagic lesions were found at the following sites: "in the left thalamus directly in the anterior tubercle, in the right thalamus also in the anterior tubercle, in the columns of the fornix, within the left cerebral peduncle, includ-

ing the red nucleus, within the mesencephalic tegmentum, the left inferior colliculus and the mammillary bodies." These findings were compatible with Wernicke's encephalopathy. Wallis *et al.*,⁶ described a case of Wernicke's encephalopathy with hypothermia being "akinetic and mute," only slightly reacting to nociceptive stimulation, forty hours after the start of thiamine therapy their patient was eating by himself and answering simple questions.

The anatomical lesions of Wernicke's encephalopathy involve the mammillary bodies symmetrically, and in various degrees the paraventricular regions of the thalamus and the hypothalamus, the periaqueductal region of the midbrain, the reticular formation of the midbrain, the inferior colliculi, the floor of the fourth ventricle and the superior vermis. The type and topography of these lesions may correspond well to the "tiny lesions of the posterior diencephalon and adjacent midbrain" held responsible for akinetic mutism."⁴

MW MANN

JD DEGOS

Department of Neurology,
CHU H. Mondor,
F-94010 Créteil, France

References

- 1 Cairns H, Oldfield RC, Pennybacker JB, Whitteridge D. Akinetic mutism with an epidermoid cyst of the 3rd ventricle. *Brain* 1941;64:273-90.
- 2 Fischbeck KH, Simon RP. Neurological manifestations of accidental hypothermia. *Ann Neurol* 1981;10:384-7.
- 3 Nielsen JM. Wernicke's encephalopathy. Correlation of sites of lesions with symptomatology. *J Nerv Ment Dis* 1953;118:429-34.
- 4 Plum F, Posner J. *Diagnosis of Stupor and Coma*. 3rd Ed. Philadelphia: FA Davis Company, 1982.
- 5 Victor M, Adams RD, Collins GH. *The Wernicke-Korsakoff-Syndrome*. Oxford: Blackwell Scientific Publications, 1971.
- 6 Wallis WE, Willoughby E, Baker P. Coma in the Wernicke-Korsakoff-syndrome. *Lancet* 1978;ii:400-1.
- 7 Wernicke C. *Lehrbuch der Gehirnkrankheiten fuer Aerzte und Studierende*. Berlin: Theodor-Fischer-Verlag, 1881;2:220-42.

Accepted 2 November 1987

Hemichorea and its denial in a case of caudate infarction diagnosed by magnetic resonance imaging

Sir: The possible cognitive functions of the basal ganglia have recently received consid-

erable attention,¹ but few case reports of cognitive deficits due to lesions localised to the basal ganglia have appeared in the literature. We report a case of denial of hemichorea in a patient with a discrete lesion in the head of the left caudate nucleus, which was diagnosed by magnetic resonance imaging (MRI).

A 51 year old right-handed man was admitted to Kawasaki Seitetsu Chiba Hospital (Chiba, Japan) on 22 September 1986, because of the sudden onset of right-sided involuntary movements 2 days previously. According to family members, the movements were not seen when the patient was asleep.

On admission, the patient was fully conscious and oriented, garrulous and seemingly happy. His wife said he had a forthright personality. Visual acuity and the visual fields on confrontation were normal; the pupils were equal and reacted normally to light. Extraocular movements were also normal. Other cranial nerve functions, limb muscle power, and muscle stretch reflexes were intact. There was some decrease of the tone of right-sided muscles and the plantar responses were flexor. Nearly continual distal choreic movements of the right arm and leg were observed without proximal ballistic movements. There was a slight decrease in pinprick sensation and vibration on the right leg but position and light touch sensation were normal. These sensory abnormalities disappeared rapidly within 3 days.

The patient was found to be totally unconcerned about the chorea. Although the involuntary movements made the playing of games, such as chess, impossible, he invited other patients to play and tried to undertake activities which were manifestly impossible. Moreover, when asked about his right limbs, he responded that they were fine or had recovered, despite the fact that violent involuntary movements were ongoing. In this respect, he showed clear signs of denial of the hemichorea.

Neither hemineglect (as determined by horizontal line bisection, copying pictures and Albert's line-crossing test) nor constructional apraxia (copying figures such as a diamond shape, a Greek cross and a cube) were found. There was no extinction on double simultaneous tactile and visual stimulation. Facial recognition, identification of overlapping figures (Poppelreuter), colour naming, limb and facial praxis to command, left-right discrimination, finger naming and simple verbal calculations were intact. His Wechsler Adult Intelligence Scale was at a normal level (Verbal IQ = 103, Performance IQ = 92 and Full Scale IQ = 110).

Computed tomography (CT) performed on 6 October 1986 (on the 17th day from onset) showed a small low density area in the head of the left caudate nucleus (approximately 4 cm rostral to the orbitomeatal line). The low density area did not extend to the posterior portion of the internal capsule or to the globus pallidus, and no other abnormalities were seen. Enhancement after contrast infusion in the region of the left caudate nucleus indicated recent infarction. Magnetic resonance imaging (Picker International Ltd., VISTR-MR) was performed with a 0.5 Tesla superconducting magnet using the inversion recovery technique (on 20 December, the 92nd day from onset). The width of the slice was 10 mm and the patient's head was positioned supine. Horizontal (fig a) and left lateral sagittal (fig b) images showed a small low intensity area in the head of the left caudate nucleus. There were no other abnormalities.

The course of the disease has been favourable. The patient was treated with haloperidol (0.75 mg, thrice daily) for three days, starting on 1 October, the 12th day from onset. The severity of the involuntary movements decreased and the haloperidol therapy was then stopped. The hemichorea gradually improved and had disappeared within six months. In contrast, the patient's denial of the hemichorea continued for three weeks from onset, but disappeared during the period when hemichorea was no longer observed.

The present case showed sudden onset of right-sided chorea and some decrease of muscular tone, but no symptoms of the pyramidal tract, such as motor paresis. Mild sensory abnormalities were observed in the right leg but rapidly disappeared and the patient was diagnosed as acute hemichorea. MRI clearly revealed a lesion localised to the head of the caudate nucleus contralateral to the involuntary movements.

Particularly noteworthy about this case was the denial of the hemichorea during the early stages of the disease. This finding is analogous to the denial of hemiplegia (anosognosia) first reported by Babinski² and indicates that anosognosia can also be found in cases of involuntary movements. Weinstein³ has previously described a case with diffuse brain tumour in which there was denial of involuntary movements, and Goldblatt *et al.*⁴ have also reported the denial of chorea in the acute stage in a case of hemichorea in which infarcts were confined to two locations in the caudate nucleus and the putamen.

Anosognosia is normally considered to be a deficit of higher cortical functions, and thought to be a right parietal sign.⁵ How-