arise through nutritional deficiency and not through any direct toxic effect of ethanol alone. A further uncommon pathological change found in demented alcoholics has been described by Morel (1939) as 'sclérose corticale laminaire alcoolique' but remains unclassified in this respect.

Alcoholic dementia should therefore be thought of as dementia due to alcoholism as opposed to dementia due to alcohol.

REFERENCES
Cole M, Turner A, Frank O, Baker H & Leevy C M (1969) *American Journal of Clinical Nutrition* **22**, 44–51
Morel F (1939) *Schweizer Archiv für Neurologie und Psychiatrie* **44**, 305
Phillips G B, Victor M, Adams R D & Davidson C S (1952) *Journal of Clinical Investigation* **31**, 859–871
Victor M, Adams R D & Collins G H (1971) The Wernicke-Korsakoff Syndrome. Blackwell Scientific, Oxford; p 42

Dr G Neale said that although alcohol might not cause a specific neurological lesion it did seem to have a direct effect on red cell and platelet production (Hines J D & Cowan D H 1970 *New England Journal of Medicine* **283**, 441; Cowan D H & Hines J D 1971 *Annals of Internal Medicine* **74**, 37). He asked if the hæmatological abnormalities in the patient had been studied in detail.

Dr Mallinson said that they had not.

Dr B I Hoffbrand said that it was difficult to get a clear picture of the incidence and associated features of alcoholic dementia from standard textbooks of either medicine or psychiatry.

Stiff Man Syndrome: Is the Lesion at Spinal Cord or Brain Stem Level

John Cobb BA MRCP
(The Maudsley Hospital, Denmark Hill, London SE5 8AZ)

C C, woman, born 11.11.46

History: Aged 6 the patient suffered from 'chickenpox encephalitis'. During childhood she was accident prone. Apart from this she was healthy until the onset of symptoms at 22. No significant medical or psychiatric history in her family. Her initial complaint was of menorrhagia associated with a change in her menstrual cycle from 28 to 14 days. Following this she started to experience feelings of panic on going out, together with increasing generalized anxiety, depression and insomnia. Despite an excellent work record over the previous five years she soon became unable to work and was admitted to a psychiatric hospital six months after the onset of symptoms. She was treated with a wide range of antidepressants and tranquillizers; modified ECT and ether abreactions failed to help her, and she remained in hospital for nearly three years.

The first attack of stiffness occurred two years after the onset of symptoms. In a typical attack she lay rigid, unable to move, with her neck in opisthotonos, back arched in extension, abdomen board-like and lower limbs fixed in extension, feet plantar flexed. Her pupils were widely dilated, responding only sluggishly to light and her pulse rose to 180/minute. Tendon reflexes were symmetrically increased and bilateral knee clonus was present. Plantar responses were flexor and both Trousseau and Chovstek reactions were negative. Noise, active or passive movements of limbs, emotional upsets or even swallowing precipitated severe painful spasms, which caused dislocation of the right humerus on three occasions. These symptoms persisted for several days but disappeared during sleep and under general anæsthesia. Oral diazepam in massive doses (20 mg 4 times daily) produced complete control of physical symptoms within three days, the improvement starting 30 minutes after the first dose. Double blind substitution of placebo rapidly led to return of symptoms. Barbiturates and to a lesser extent phenothiazines were also effective in reducing stiffness, but mephenesin (up to 1.5 g 4 times daily) and baclofen (15 mg 4 times daily) were without significant effect. To maintain control of the physical symptoms, the daily dose of diazepam has had to be increased to 180 mg. Despite this high dose her conscious level is unimpaired. Though able to leave hospital she remains anxious and depressed, with a moderate fear of going out alone. The menstrual abnormality persists and insomnia is still a severe problem.

Investigations: Hb 14.1 g/100 ml. WBC 7600/mm^3. ESR 8 mm in 1 hour (Westergren). Urea 17, sodium 143, potassium 4.2, chloride 103, calcium 10, PO_4 3.2, blood sugar 84 mg/100 ml (all during an attack of stiffness). Fasting plasma insulin 54 μu/ml (Searle immunoassay normal less than 25 μu/ml). Simultaneous blood sugar 82 mg/100 ml. Glucose tolerance test normal. EEG normal. EMG: 'continuous motor unit activity to a similar degree in both left triceps and left biceps. Attempts at active or passive movements produced generalized muscle spasm'. Urine: no reducing substances.

Discussion

Moersch & Woltman (1956) described a condition characterized by extraordinary muscle stiffness which they called 'stiff man syndrome'. Gordon *et al.* (1967) surveyed the world literature and established the definitive criteria which distinguish this unique syndrome from other causes of muscle stiffness. Most workers believe that the lesion lies at spinal cord level, either in the inhibitory internuncial neurones (Renshaw loop)

or in the gamma motor neurones of the intrafusal feedback loop (Gordon *et al.* 1967). However, a lesion at this level would fail to account for the metabolic and psychiatric abnormalities noted in many cases. This case is presented, not only because this is a typical example of a rare syndrome, but also because the psychiatric symptoms and some of the physical signs are better explained by postulating a lesion at brain stem level rather than in the spinal cord.

The clinical picture and EMG findings are typical of the stiff man syndrome (Gordon *et al.* 1967). It was not possible to check the effects of myoneural blocking agents or chemical block of peripheral nerves, but all the other major criteria established by Gordon *et al.* were satisfied. There are several reasons for postulating a brain stem rather than a spinal cord lesion. (1) Psychiatric symptoms preceded the onset of stiffness by an unusually long period and persisted after the stiffness had been controlled. It is difficult to dismiss these symptoms as a secondary reaction to muscular discomfort. Either the psychiatric state must be seen as a separate condition or alternatively as a primary part of the syndrome. (2) The severe insomnia and remarkable insensitivity to sedatives raises the possibility of dysfunction of the reticular formation. (3) The pupillary reaction and clonus observed during an attack of stiffness are difficult to reconcile with a spinal lesion. (4) The menstrual abnormality and raised fasting serum insulin might be explained on the basis of pituitary disorder. All these points are speculative but suggest areas in which future research on this rare but intriguing syndrome might be profitable.

Conclusion

This is a typical case of stiff man syndrome, a rare but treatable condition. Clinical features provide tentative evidence that the lesion is at brain stem level rather than at spinal cord level, as previously suggested.

REFERENCES

Gordon E E, Januszko D M & Kaufman L (1967) *American Journal of Medicine* **42**, 582–599

Moersch F P & Woltman H W (1956) *Proceedings of the Staff Meetings of the Mayo Clinic* **31**, 421–427

Dr G Neale said that the diagnosis of stiff man syndrome shortly after the onset of the illness was not easy. The symptoms, the autonomic disturbances and the response to diazepam were very similar to those seen in patients with chronic tetanus and indeed this had been one working diagnosis in some patients in whom the progress of the disease had led subsequently to a diagnosis of stiff man syndrome. Conversely, as shown by Gordon *et al.* (Gordon E E, Januszko D M & Kaufman L 1967 *American Journal of Medicine* **42**, 582), some reported cases of stiff man syndrome were probably incorrectly labelled with chronic recurrent tetanus in the partially immunized subject as the most commonly missed diagnosis.

Dr P Rudge said that surely the lesion in this patient was at a higher level than the Renshaw cell. Her rigidity, impassive facies and autonomic disturbances suggested a diencephalic abnormality involving the upper reticular formation and basal ganglia. Such a lesion could possibly explain the abnormal insulin levels during the glucose tolerance test by a disturbance of the hypothalamic pituitary axis causing, for example, growth hormone release. It would also explain the abnormalities of sleep these patients had. It might be useful to make a continuous EEG record during sleep, to see if she had a normal pattern of REM sleep, and to measure pituitary hormone levels throughout the period.

Depressive Illness and Hyperparathyroidism

Peter Noble MD MRCP

(Maudsley Hospital, London SE5 and Hammersmith Hospital, London W12)

Woman, aged 53

History: Born and educated in Denmark. Secondary education and commercial training were followed by secretarial work until her marriage at 35. Her husband, a successful business man, had 2 children by a previous marriage. She was more shy and anxious than average but seemed to have had a happy social and family life. There were no clear environmental precipitants for her illnesses.

Hypertension was diagnosed and treated with reserpine 0.025 mg daily for 4 months from October 1971. In December 1971 she developed a severe depressive illness which responded to 5 electropexies (ECT). Trimipramine 75 mg daily was prescribed until March 1973.

Present illness: In May 1973 she became apathetic and retarded. She was treated with opipramol up to 150 mg daily for four weeks and received 5 ECT without improvement. Dehydration and urinary and fæcal incontinence developed. Hypercalcæmia was discovered and the patient was transferred to the Hammersmith Hospital on 29.8.73. A parathyroid adenoma was removed on 19.9.73. There was no improvement in her mental state and she was transferred to the Maudsley Hospital on 5.10.73.

On examination: Marked apathy, psychomotor retardation and complete absence of spontaneous talk. She was unable to join in ward activities, was pessimistic about the future and complained of feeling 'flat' and 'dead inside'. Concentration was poor: memory and orientation were normal.