SIX CASES OF CEREBELLAR DEGENERATION ASSOCIATED WITH CHRONIC ALCOHOLISM

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A well-recognized symptom of acute alcoholism is a temporary interference with coordination. This usually persists for some hours or days after cessation of drinking, but may occasionally persist for a week or more, and has been attributed to the 'toxic' effect of alcohol on the cerebellum. Until fairly recently it was not generally appreciated that irreversible cerebellar degeneration might result from chronic alcoholism. (Indeed it is becoming more and more apparent that alcohol can be associated with damage to any part of the nervous system.) During the last 2 years there have been several reports of cases of chronic cerebellar degeneration associated with chronic alcoholism. The most impressive of these is a report by Victor, Adams and Mancall[1] on the clinical details of 50 cases and autopsy details of 11 cases. These authors were struck by the uniformity of the clinical picture. Almost invariably the gait and lower limbs were grossly affected; the upper limbs were often not affected and dysarthria and nystagmus were uncommon.

'The essential pathological change in 11 autopsied cases was a degeneration of all the neurocellular elements (but especially of the Purkinje cells) of the cerebellar cortex. This degeneration was strikingly selective in that it was restricted to the anterior and superior aspects of the vermis and the hemispheres. The olivary nuclei were almost always involved; the fastigial, globose, emboliform

and vestibular nuclei were less consistently affected. The dentate nuclei, cerebellar white matter and peduncles, the spinocerebellar tracts and other brain-stem nuclei were essentially unaltered'.[1] The portion of the cerebellum which lies anterior to the primary fissure is generally regarded as the 'leg area' in experimental animals and this explains the predominant involvement of lower limbs seen clinically.

From August 1959 to May 1960, 6 chronic alcoholic patients with cerebellar degeneration were admitted to Groote Schuur Hospital. The clinical details are presented.

CASE REPORTS

1. W.E. (European male), aged 49 years

This patient was admitted to the ward in August 1959. He stated that he had been unsteady on his feet since 1955 or 1956. The onset of the unsteadiness was slow but had been steadily progressive, so that he had been afraid to walk in the dark during the last few months. He had to use his arms to pull himself up when climbing stairs.

On examination his general condition was good. He was normotensive. The liver was 3 fingerbreadths below the costal margin, and firm but not tender on palpation. The pupils were small and slightly irregular, and reacted sluggishly to light.

The upper limbs were normal in power, tone, reflexes, sensation and coordination.

In the lower limbs there was no motor weakness but the tone was diminished and the ankle jerks were only just elicited with reinforcement. There was some patchy diminution of light touch and diminution of pain sensation over the dorsum of the feet. Calf-muscle tenderness was not increased. There was gross heel-knee ataxia, marked ataxia on attempting to walk along a straight line and marked Rombergism. The cerebrospinal-fluid chemistry and serology were normal. Blood serology was normal. The patient was given a full diet, 'plebex' intramuscularly, and later vitamin-B compound by mouth. He was discharged 4 weeks after admission and on discharge could walk along a straight line, his heel-knee ataxia was less marked, and Romberg's test was negative.

2. C.D. (Coloured male), aged 35 years

This man was admitted in August 1959, complaining that for a year he had had difficulty in walking. He could not analyse this difficulty precisely but said that his feet felt 'heavy' and he was not quite sure where they were on the ground. This difficulty with walking had progressed to such an extent that for 3 weeks before admission he had been bedridden. For 6 months his speech had been indistinct. For 2 months he had had a dull pain in the lower limbs.

On examination his general condition was good. Blood pressure was 150/100 mm.Hg. The liver was 2 fingerbreadths below the costal margin and firm to palpation. Mild dysarthria was present. The upper limbs were normal apart from mild incoordination in the finger-nose test. In the lower limbs the knee and ankle jerks were absent and the calf muscles were tender. There was gross heel-knee ataxia and his gait was ataxic. (Incidental findings were a positive blood serology and a slightly widened aorta. He was given 36 million units of penicillin while in the ward.)

He was discharged after 3 weeks with his condition unchanged. He was re-admitted in January 1960, and stated that he was walking better, his speech was more distinct and his calves were no longer tender. He had cut down his drinking to some extent. On examination he had minimal dysarthria. The right upper limb showed a coarse tremor at rest but coordination was probably normal. There was gross heel-knee ataxia and heel-toe incoordination was present when walking.

3. N.S. (European female), aged 39 years

This patient was incapable of giving an adequate account of herself and no relatives or friends could be interviewed. In July 1959 she was admitted to a hospital with a diagnosis of Korsakoff's syndrome, peripheral neuritis and Wernicke's encephalopathy. In August she was transferred to a convalescent home and was reported at that time to be normally ambulant, helpful and cooperative, but vague about her past. While at the convalescent home she developed increasing tremor and became unable to walk or to feed herself. She was admitted to Groote Schuur Hospital in December 1959.

On examination she was in good general condition. She cooperated well in examination and her attention was sustained. There was no overt evidence of intellectual deterioration and tests of arithmetical ability were well done. However, the simplest questions demonstrated gross memory impairment, especially for recent events and all details of her illness. There was a marked nodding tremor of the head and at rest an occasional and variable tremor was present affecting the thumb or a finger; it consisted of a slight abduction-adduction movement. Voluntary movements accentuated this tremor and it spread so that a pronation-supination movement of the forearm was noted in addition to a tremor of the leg when carrying out the heel-knee test.

There was moderate dysarthria with a 'scanning' element to the speech. Nystagmus was not present, there was no cranial-nerve defect, and power, reflexes and sensation were normal. Tests of cerebellar function, however, demonstrated marked ataxia and tremor in the finger-nose test but fairly good performance of rapidly alternating movements. In the lower limbs there was no heel-knee ataxia and toe-object touching was normal. Stance and gait were grossly abnormal. The patient stood on a wide base with knees hyper-extended and involuntarily sought support.

The cerebrospinal fluid and the serum proteins were normal. Electro-encephalography showed generally low-voltage wires with irregular fast frequencies and some random slow α-activity.

She was discharged in April 1960 and, despite intensive vitamin therapy and physiotherapy, had made no essential progress.

4. E.S. (African female), aged 58 years

This patient had noticed increasing unsteadiness of gait for 10 months. For 2 months before admission she was unable to walk without support.

On examination her general condition was good but she was moderately hypertensive (blood pressure 170/105 mm.Hg).

The only neurological abnormalities were a grossly ataxic gait, gross heel-knee ataxia and intention tremor of the right leg on toe-object testing, in addition to mild peripheral neuritis (sluggish ankle jerks and increased calf-muscle tenderness). Liver-function tests and serum proteins were normal. Cerebrospinal-fluid chemistry and serology were normal. The electro-encephalogram was normal. Chest X-ray examination showed a calcified tuberculous focus in the left lower lobe.

5. A.L. (European male), aged 34 years

This patient was first admitted to a medical ward on 6 January 1960 and was quite incapable of giving any account of himself. His wife was not very informative but said that her husband had suffered from epileptic seizures 'for some years'. Two days before admission he began to have about 4 seizures a day and since then had been confused.

On admission he was confused. His general condition was poor and he had marked palmar erythema. Both feet were cold and red. He had no cranial-nerve defect and particularly no dysarthria. In the upper limbs the finger-nose test was passably well performed but rapidly alternating movements were poorly performed. In the lower limbs power was normal, but neither knee nor ankle jerks could be obtained and the calf muscles were tender. In addition there was moderate heel-knee ataxia and gross ataxia on walking so that he was unable to walk without support.

When re-examined in July 1960, there was no real change in his condition. It was then apparent that he had gross intellectual deterioration. In addition a tremor was seen involving the upper limbs (right more than left), consisting of a rapid distal pronation-supination movement at rest, resembling a Parkinsonian tremor. Serum proteins, liver-function tests and cerebrospinal fluid were normal.

6. K.A. (Coloured male), aged 35 years

This patient was admitted to a medical ward in May 1960. He was unable to give any account of himself — he was

confused and disorientated, and had gross memory loss.

His cranial nerves and upper limbs were normal. He had a mild peripheral neuritis (knee and ankle jerks barely obtainable, calf muscles tender and soles hyperaesthetic) with gross heel-knee ataxia. He was unable to walk without support.

DISCUSSION

These 6 patients (2 of whom — N.S. and E.S. — were women) were between the ages of 34 and 58 years. All were known to be severely alcoholic. The only time this was questioned was in the case of E.S., the African female, who admitted consuming large amounts of home-brewed beer. This is usually regarded as having a low alcohol content unless adulterated. Four of the patients had been drinking shortly before their admission to hospital but N.S. had been hospitalized, and was thus presumably without alcohol, for 6 months before admission; and the wife of A.L. stated that he had not had anything to drink for several months because he felt too ill.

All the patients were in fairly good general condition and looked well-nourished. Two (W.E. and C.D.), had enlarged firm livers, but all special investigations, e.g. liver-function tests, serum proteins, etc. were normal.

All patients had normal chest X-rays and no evidence of malignant disease.

All the patients had other neurological involvement in addition to cerebellar degeneration. All had evidence of peripheral nerve disease; A.L. and K.A. were grossly confused, had memory loss, and could not give any account of themselves; while N.S. had a remarkable and gross defect of memory extending back for several years, but no other evidence of intellectual deterioration. The cerebrospinal fluid was normal in 5 of the cases in whom it was examined.

Electro-encephalograms were performed in 2 patients — E.S., in whom it was normal, and N.S., where it was reported as showing generally low voltage with irregular fast frequencies and some random slow α-activity.

The cerebellar signs gave a picture, in 5 patients, conforming closely to the cases reported by Victor *et al.*[1] All 5 patients were grossly ataxic and had marked heel-knee ataxia. In 3 cases (W.E., E.S. and K.A.) this was the only clinical evidence of cerebellar defect. C.D. had mild incoordination of the upper limbs and slight dysarthria, while A.L. had difficulty in performing rapidly alternating movements with the upper limbs though the finger-nose test was normal in his case. Therefore, like the cases of Victor *et al.*,[1] there was gross involvement of the lower limbs with minimal or no upper limb defect. One patient, N.S., was unusual in that she had fairly marked dysarthria and marked ataxia and tremor in the finger-nose test, but in the lower limbs, despite gross ataxia, there was no heel-knee ataxia and toe-object testing was normal. Only one patient, W.E., showed any clinical improvement of the cerebellar signs. The other patients, despite weeks or even months of hospitalization with intensive vitamin therapy and physiotherapy, were eventually discharged essentially unchanged. In 2 cases (N.S. and A.L.), there was a tremor similar to that seen in basal ganglion disease which could not be explained by cerebellar cortical degeneration. The precise pathogenesis of the cerebellar degeneration remains a mystery.

SUMMARY

The clinical picture of chronic cerebellar degeneration in 6 chronic alcoholic patients is described in detail.

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REFERENCE

1. Victor, M., Adams, R. D. and Mancall, E. L. (1959): A.M.A. Arch. Neurol., 1, 579.