an altered level of consciousness, for “all patients were awake throughout the investigations and were checked repeatedly to see that they were capable of verbal communication”. Subsequently, Lavin et al described similar observations in another group of eight patients with Huntington’s disease, in all of whom chorea disappeared for at least an hour within about half-an-hour of the insulin injection. Such a dramatic effect on chorea might provide some clue as to the pathophysiology of that movement disorder, so we have repeated the study concentrating on the effect of insulin-induced hypoglycaemia on the chorea.

Five patients with Huntington’s disease (four males and one female; aged 30 to 70 years; with disease duration from 2 to 13 years; four on no drugs and one on tetrabenazine 25 mg three times daily) with obvious chorea were studied. After an overnight fast, blood was withdrawn for glucose estimation, and insulin (0.1 mg/kg) was injected into the opposite arm. Blood sugar and clinical response were measured every 10 min for 60 min, and then every 20 min for a further 60 min. The severity of chorea was rated using a specially designed scale described in detail elsewhere. In addition the number of choreic movements occurring at rest in one selected region, such as an eye, finger or toe depending on the individual patient, was counted over a 60 sec period. Blood sugar fell below 2.0 mmol/l and symptoms and/or signs of hypoglycaemia developed in all subjects. However, the intensity of chorea did not alter. Three subjects fell asleep during the test, and chorea disappeared in two, but on arousal their chorea was of the same severity as before insulin. Unfortunately, insulin-induced hypoglycaemia had no effect on chorea in our patients.

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References


Periodic alternating nystagmus in a case of hereditary ataxia and its treatment with baclofen

Sir: I describe below a patient with a dominantly inherited cerebellar degeneration who presented with oscillopsia, and was found to have periodic alternating nystagmus. His symptoms were virtually abolished following treatment with baclofen, which confirms a recent finding in two out of three cases of periodic alternating nystagmus similarly treated. Periodic alternating nystagmus itself is a rare and extraordinary form of spontaneous nystagmus. It is a horizontal, usually jerk nystagmus which changes direction periodically. Less than a hundred cases are to be found in the literature in association with a considerable variety of disorders including multiple sclerosis, posterior fossa malformations and tumours, phenytoin intoxication and neurosyphilis. One previous case has been reported in a hereditary ataxia (Friedreich's ataxia), but the veracity of the diagnosis in that case has been questioned.

A 34-year-old salesman presented with a four year history of increasingly troublesome oscillopsia. His father and grandfather had both developed ataxia in middle life. The former is living, having been investigated elsewhere, and a diagnosis of hereditary ataxia made; he has nystagmus, but details are not known. No other family members are as yet affected. On examination, visual and alternating nystagmus were noted, but no other abnormalities. Routine blood count and biochemistry were normal. Acanthocytes were looked for but none seen. Serological tests for syphilis were negative. Skull radiographs were normal but a CT scan showed marked brain stem and cerebellar atrophy. Nerve conduction studies and visual evoked responses were within normal limits. The upper trace in the figure is a continuous record of the patient's periodic alternating nystagmus with eyes in the primary position. The cycle length is 182 seconds with a left beating phase of 80 seconds, a right beating phase of 76 seconds and two null phases of 12 and 14 seconds respectively. The second trace shows a series of saccades 30° to either side of the primary position, recorded at a lower gain. At the start of the trace the null position is at 30° left, as it shifts to the primary position first degree nystagmus to the left appears and increases in amplitude. It has been commented previously that the nystagmus can be conceptualised as resulting from periodic shifts of the null zone. Other features were also similar to most previously reported cases (for example the three analysed in detail by Baloh et al). The spontaneous nystagmus was superimposed on saccades and smooth pursuit. Optokinetic nystagmus could be produced only during null phase or when the stimulus was moving in the same direction as the slow phase of the nystagmus.

Traces three and four were recorded in a similar manner whilst the patient was taking baclofen 10 mg thrice daily. Spontaneous nystagmus in the primary position has been abolished, first degree nystagmus still occurs during saccadic eye movements to right and left but there is no longer any shift of the null position.

Oscillopsia is common in periodic alternating nystagmus. It is particularly troublesome because, for most of the cycle, nystagmus occurs in the primary position. This patient was rendered asymptomatic for most activities whilst taking baclofen, because this feature was abolished.

Periodic alternating nystagmus is uncommon and easily missed. The present case illustrates that whenever the underlying disorder, it is worthwhile looking specifically for the condition in any patient complaining of oscillopsia as it appears to be one of the few eye movement disorders for which effective treatment is available.

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References

Painless cauda equina schwannoma simulating Charcot-Marie-Tooth disease

Sir: Pain is almost always the principal symptom of patients who have tumours of the cauda equina. Painless tumours at this site, although rare, do occur and should be considered in the differential diagnosis of pain free patients presenting with progressive weakness in the lower limbs. The following case report demonstrates this point.

A man, aged 38 years, was admitted with 10 years' history of progressive weakness of the legs, the left being more severely affected. There was no history of any pain or paresthesia in his back or legs. Other symptoms included a recent constant feeling of urgency, occasional nocturia, constipation, and weak penile erection. Previously he was admitted to another hospital because of a left ankle sprain, leg weakness, and a gait disturbance, at which time a diagnosis of peroneal muscular atrophy was made. The abnormal neurological signs were confined to the lower limbs. Moderate to severe weakness and wasting, especially of hip extensors, foot dorsiflexors, extensors of toes, foot eveters and invertors, were present with bilateral foot drop. Knee jerks, ankle jerks, and patellar responses were normal. There were no sensory changes. The rectal sphincter was hypertonic. Straight leg raising to 90° produced no pain or discomfort. Lumbar spine radiographs were normal. Nerve conduction studies with standard techniques showed normal maximum conduction velocities in right median, ulnar, tibial, and sural nerves, and borderline conduction velocities in left tibial nerve. Supramaximal percutaneous stimulation of the peroneal nerves produced no muscle action potential. Needle electromyography showed extensive denervation potentials in glutei maximii and in the muscles innervated by both sciatic nerves but no abnormality in paraspinal muscles. Myelography revealed a mass at the cauda equina region. Spinal fluid protein was elevated (1·28 g/l). Urodynamic studies were consistent with a spastic neurogenic bladder. Laminectomy of D12 and L1 vertebrae was performed and a large Schwannoma was removed totally.

Cauda equina tumours are rare. Occasional cases present with unusual clinical features such as pseudoclaudication, foot ulceration, subarachnoid haemorrhage, papilloedema, and sensory ataxia, but it is generally accepted that pain in the back and lower limbs is nearly always the most important and early presenting symptom. Pain may be of sudden or gradual onset, usually tends to become constant, and characteristically worsens at night. Allen and Spiller reviewed the literature of cauda equina tumours and both found only one case of cauda equina compression with no pain, reported by Volhard. Campbell could find clinical details of only two cases of painless tumour of the cauda equina and presented one of his own. In a series of 20 patients from the National Hospital, Queen Square, only one had pain and presented with left leg weakness for seven years; but finally he developed severe lumbar pain. In an Oxford series of 70 patients, six complained of painless progressive weakness of legs; only three had bilateral leg weakness, and pain was the presenting symptom in 31 out of 34 patients with neurofibromas of the lumbar-sacral region.

In sum, a case is reported of a patient with a Schwannoma of the cauda equina who carried the diagnosis of Charcot-Marie-Tooth for 10 years. There was slowly progressive motor dysfunction without pain or sensory loss during this time. Electromyography excluded Charcot-Marie-Tooth disease, and indicated a lesion of the cauda equina and, therefore, manded myelography. This emphasises the clinical significance of proper electromyographic study in patients with progressive neuromuscular disease.

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