Cervical nystagmus due to loss of cerebellar inhibition on the cervico-ocular reflex: a case report

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SUMMARY Studies of the cervico-ocular reflex and the vestibulo-ocular reflex have been carried out separately and in combination on a patient with gait ataxia due to a cerebellar tumour. With the head fixed in space, body rotation to the right (left neck torsion) induced marked nystagmus to the left in darkness. Vestibulo-ocular responses to sinusoidal rotation were symmetrical while the neck was immobilised and asymmetric when it moved freely. It is suggested that the cervical nystagmus seen in this case was the result of removal of cerebellar inhibition upon the cervico-ocular reflex and that an abnormal interaction of cervical and vestibular inputs could have played a role in the patient's unsteadiness.

While clinical and experimental studies have provided abundant evidence of the inhibitory role played by the cerebellum in the vestibulo-ocular reflex, much less is known about its influence upon the cervico-ocular reflex. This stems in part from the difficulty in eliciting the reflex in normal animals and humans. In addition, although in certain clinical conditions nystagmus can be elicited on neck torsion, it is usually difficult to exclude a vascular aetiology. The present paper is concerned with a unique case in which a nystagmus of undoubted cervical origin has been demonstrated, due, in all probability, to a derangement of the central connections.

Case report

Two weeks before admission a 58-year-old male pipe smoker developed occipital headache and difficulty in walking. Initially the latter was only evident on turning when he would sway from side to side. Walking in a straight line was unimpaired. This symptom gradually increased, however, and at the time of his admission he staggered markedly. A few days before admission he developed very mild hand clumsiness and slurred speech. The headaches had also been increasing and he was anorexic and vomited on one occasion.

General examination was unremarkable; blood pressure was 140/80 mm Hg. Neurological examination disclosed a broad based stance and marked gait ataxia; he was especially unsteady on turning while walking. Cranial nerves, fundi, sensation and muscle power were normal apart from diminished pin-prick sensation on the feet. There was a moderate lower limb ataxia but upper limbs were considered normal. In the lower limbs, deep reflexes were brisk and tone was thought to be marginally increased but plantar and cutaneous abdominal reflexes were normal. There was a moderate dysarthria. On oculomotor examination no palsies were found. No spontaneous nystagmus on primary position of gaze but gaze evoked nystagmus to right, left and upwards was elicited. Lying down, without neck torsion, either with right or left ear down, produced a non-fatiguing nystagmus beating towards the upper ear, without latent period or vertiginous sensation. Extreme neck torsion to right and left in the sitting position did not induced any nystagmus or vertigo. Smooth pursuit was broken up (saccadic) in all directions. Saccades were slightly hypometric, but seemed to have normal speed.

A CT scan showed a cerebellar mass involving the anterior vermis, paramedian region and partial hemisphere on the right (fig 1). An occipital craniectomy was carried out; histological diagnosis of undifferentiated adenocarcinoma was made. A chest radiograph revealed a lung tumour thought to be the origin of the cerebellar metastasis.

Pre-operative investigations

Cervico-ocular reflex (COR) In order to examine the role of the neck reflexes in isolation it is necessary to rotate the trunk while maintaining the head immobile. To this end the subject sat in a rotating chair which could be moved manually. In this position he was required to bite on a
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Cervico-ocular reflex (VOR) The cervico-ocular reflex was studied under conditions of both passive and active head movement. In the former, the patient with head firmly secured to the chair was rotated sinusoidally in total darkness at a frequency of 0.3 Hz over a total amplitude of ± 25°. Both chair displacement and eye movements were recorded simultaneously. In the case of active head movements the subject, again in total darkness, was instructed to move his head sinusoidally at 0.3 Hz in time with a metronome. The resultant ocular response thus combined both vestibular and cervical inputs. Head movement was monitored by means of a light helmet attached to a low torque potentiometer.

Results

Cervico-ocular reflex
(a) Ramp stimuli Rotation of the trunk, say to the right, resulted effectively in relative movement of the head to the left and for convenience will be referred to as HL; likewise with trunk to left as HR.

All trunk movements to right (HL) irrespective of the starting position, evoked a marked saw-tooth type nystagmus to the left with an exponential decay of the slow phase component reminiscent of gaze evoked nystagmus of cerebellar origin. On cessation of movement, presentation of the target light resulted in an eye movement of about 15° to the right indicative of a sustained tonic deviation of the eyes to the left. A typical example is shown in fig 2.

Fig 1 Enhanced CT scan at the level of the superior aspect of the posterior fossa showing rounded abnormal enhancement in the mid line and right cerebellar hemisphere associated with a small amount of oedema. The fourth ventricle was distorted and displaced to the left but the rest of the ventricular system was only mildly dilated.

Fig 2 Cervico-ocular reflex—ramp stimuli. Eye movement recordings in darkness with head fixed and trunk rotated. Upper tracing shows minimal nystagmic response to right on trunk rotation from right to left (neck torsion to right). Lower tracing shows marked left beating nystagmus induced by trunk rotation from left to right (neck torsion to left). Vertical arrows indicate presentation of target light in primary gaze position. In the lower tracing this reveals a marked tonic deviation of the eyes to the left in the absence of optic fixation.
In contrast, the responses to trunk movement to left (HR) were weak and variable although in six out of ten trials a low amplitude nystagmus to the right appeared which was not dissimilar to responses found in normal subjects. Defining gain as the ratio of peak slow phase velocity to peak head velocity, a clear asymmetry of the COR is apparent with (HL) gain of 1.40 ± 0-65 and a (HR) gain of 0.10 ± 0.10.

(b) Sinusoidal stimuli As shown in fig 3, eye movement responses to sinusoidal movement of the trunk were remarkable in that peak displacement to the right (HL) consistently appeared to trigger large amplitude rapid components to the left followed, in turn, by a slow phase to the right which was most obvious at higher frequencies. Peak displacement to the left (HR) elicited small amplitude saccadic movements to the right.

Discussion

The main finding of importance which has emerged from these studies is the appearance of a well-marked nystagmus to the left on trunk movement to the right (HL) with head restrained (fig 2). The nystagmus appears initially during the trunk movement and persists for some time after cessation of movement. Adopting similar test procedures a cervical nystagmus of this kind can be readily demonstrated in patients with absent labyrinthine function. Indeed it is maintained that the resultant gain in the COR plays a compensatory role for the loss of the Vestibulo-ocular reflex (VOR). On the available evidence, however, it would seem unlikely that the peripheral vestibular apparatus of our patient was in any way impaired. There remains the possibility of a vascular aetiology. This can certainly be called in to account for the so-called cervical nystagmus/vertigo resulting, for example from whiplash injury or vertebro-basilar insufficiency, in which mechanical compression of the vertebral arteries during neck movements can affect blood supply of the labyrinth and the brain stem.

If this were the case, however, it is to be expected that it would be most marked on extreme neck torsion; instead all ramp movements were equally effective in eliciting nystagmus whether the movement was from mid-line to HL or HR to mid-line. In the circumstances our conclusion is that the nystagmus is the direct out-
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come of the cerebellar lesion which in turn implies that the COR is under the inhibitory control of the cerebellum. This is perhaps not surprising since it is well established that the vestibulo-cerebellum exerts a similar inhibitory effect upon the VOR.1 According to the CT scan the vestibulo-cerebellum would almost certainly have been spared with the lesion mostly involving anterior and rostral parts of the posterior lobes. These areas, however, are known to receive strong proprioceptive projections including those from the cervical receptors.12-14

Apart from the typical gait ataxia which results from lesions in this region our findings suggest that a new factor which needs to be taken into account is abnormal interaction of the cervical and vestibular inputs. The effect of this is best illustrated in the abnormalities found during active head movements exemplified in fig 5. These in turn could give rise to subtle postural disorders such as the imbalance experienced by our patient on turning while walking.

Although to the best of our knowledge these findings are unique, it is perhaps worth stressing that they have come about largely by the chance application of somewhat sophisticated test procedures to a particular patient during the course of a more general enquiry into the role of cervical proprioception. Ill-defined postural abnormalities related to head movement are not uncommon in patients with central disorders and a more systematic investigation of this kind might well reveal that associated derangements of the kind described here are the rule rather than the exception.

References