treatment and plasmapheresis are effective in preserving sensory nerve potentials and motor function.

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Contraversive visual tilt illusion associated with a cerebellar infarction

Visual tilt illusion consists of an abnormal perception of the environment, which seems to be rotated at a variable angle without any change in the patient’s mirror image or other characteristics of the objects. It is sometimes associated with other postural and ocular tilt effects. It can be secondary to disturbances in the peripheral or central vestibular pathways.1 2 Previous reports suggest that cerebellar injuries could also cause it1 but this has not been documented before. We report a case of visual tilt illusion probably associated with an isolated cerebellar lesion, studied with CT and MRI.

A 56 year old man with hypertension and hypercholesterolaemia had a sudden attack of vertigo, nausea and vomiting, which seemed to worsen on head and body tilt to his left that worsened when he was asked to close his eyes and stand upright. There was no skew deviation or other ocular motor disorders. Pupillary photographs were not taken, so that ocular torsion could not be reliably assessed. There were no other alterations on neurological examination. Two weeks later the patient was asymptomatic. Brain CT and MRI showed a right hemispheric cerebellar region, suggesting an ischaemic infarction (figure). No brainstem or other cerebellar correlations were found.

Visual tilt illusion has been described in unilateral peripheral vestibular lesions; in brainstem injuries, typically in the Wallenberg syndrome, in other medullary and mesencephalic lesions, and in thalamic and parietooccipital disorders.1-4 One of the most frequent conditions associated with visual tilt illusion are vascular lesions.1 As far as we know, there are no reports on documented isolated cerebellar injuries associated with the illusion.

Physiologically, the vestibular pathways make contact with the ocular motor system, the spinal cord, and the vestibular cortex, contributing to the stabilisation of posture and perception of verticality and self motion.1-5 The tonic bilateral vestibular input builds up the actual central vestibular tone in the three major planes: horizontal or “yaw”, sagittal or “pitch”, frontal and “roll”.6-8 It seems that central pathways that mediate vestibular function in either of the three planes travel independently of each other, so that a specific lesion could cause a disorder restricted to one of them.6-8 The vestibular tone in the frontal or “roll” plane allows a correct perceptual, ocular, and postural alignment to the “gravitational vertical”, an imbalance in this tone causes a tilt in the frontal time with alteration in perception of verticality, head and body posture, misalignment of visual axes, or ocular torsion.6-8 Patients perceive the surroundings and their body as if they were tilted in the opposite direction to what the CNS erroneously computes as being vertical and try to adjust the visual objects and posture to it. Dieterich and Brandt showed that an alteration in the perceived verticality is not just the sensory consequence of the rotation of the eyes, as they can appear separately and are not proportional in degree.9 Furthermore, it is possible that not all the effects of tilt occur in one patient, and the perceptual disorder itself is the most sensitive sign of a vestibular tone imbalance in the frontal plane.7 Brainstem structures that mediate the vestibular tone in the “roll” plane include the vestibular nucleus and the interstitial nucleus of Cajal—perhaps the most rostral structure related to the control of vertical and torsional head and eye position. Both are connected by the medial longitudinal fasciculus, which crosses the midline in the pons.2 Visual vertical tilt is, then, ipsiversive to peripheral or pontomedullary lesions and contraversive to pontomesencephalic lesions and, in both cases, is usually associated with other tilt effects; in most rostral lesions it may be either ipsiversive or contraversive and is usually isolated.10 The role of the vestibular cerebellar structures with respect to the control of subjective verticality is not well known at the moment.2

Our patient’s clinical findings suggest that he had an inclination of the internal representation of the gravitational force to his left and he tried to adjust both visual objects and posture to what he perceived as being vertical. It would have been interesting to assess whether there was ocular torsion, to define his clinical setting more exactly, but it makes no difference to interpretation as ocular torsion can be associated or not with perceptual or other tilt effects.1 Our patient showed a right hemispheric cerebellar ischaemic lesion, in a territory dependent on the posterior inferior cerebellar artery (PICA), with no mass effect and no brainstem or other alterations on MRI. The contraversive postural and visual tilt was contraversive to the lesion. It is possible that an additional subacute medullary lesion in the distribution of the PICA, not evident with clinical and imaging studies, produced the tilt effects in this case, because this type of infratentorial lesions supplies both brainstem and cerebellum and it is very difficult to differentiate the effects of cerebellar and brainstem lesions.2 But the tilt should then be ipsiversive, not contraversive, to the hypothecial lesion. Therefore it is not likely that an associated medulldary ischaemia could cause the tilt effects in our patient. A mesencephalic injury could cause this clinical picture but there were no other upper brainstem symptoms and MRI was normal at this level. A supratentorial disorder is unlikely because there were no MRI alterations and there were associated postural tilt effects. In this patient, we think that cerebel­lar dysfunction could be responsible for the tilt effects.

The present report confirms a previously hypothesised role for the cerebellar structures in the control of perception of verticality, and may contribute to a better knowledge of the pathophysiology and the topographic diagnosis of the central vestibular syndromes.

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Low striatal D2 receptor binding as assessed by [*3H]IBVM SPECT in patients with writer’s cramp

Writer’s cramp is a form of idiopathic focal task specific dystonia. In accord with other studies on idiopathic and symptomatic dystonia, Tempel and Perlmutter suggested the presence of an abnormal striatohyalamocor-

tical drive in writer’s cramp.1 In view of the

Axial T2 weighted (SE 2000/80) MRI shows an increased signal intensity in the right hemispheric cerebellar region without mass effect, corresponding to an ischaemic infarction in the territory of the posteroninferior cerebellar artery.
Letters to the Editor

Demographic data, [123I] IBZM SPECT, and bradykinin gesture pegboard test in patients with writer's cramp and controls

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Duration of disease (yr)</th>
<th>[123I] IBZM SPECT</th>
<th>Pegboard (s)</th>
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<tr>
<td>Writer's cramp (n = 10)†</td>
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<td>8-6 (3-9)</td>
<td>1-56 (0-10)***</td>
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<td>10-0 (1-4)</td>
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Values in parentheses are SD. ***P < 0.001 vs controls.

†Four patients had simple writer's cramp and six had dystonic writer's cramp.

Possible involvement of dopaminergic striatal receptors in dystonia, we measured the availability of striatal D2 receptors in patients with writer's cramp using [123I] IBZM SPECT.

Ten consecutive right-handed patients (eight male and two female) were classified into two groups: one with writer's cramp and the other with dystonic writer's cramp depending on whether or not the symptoms appeared only during writing. None of the patients had been treated with neuroleptic, dopaminergic, or anticholinergic drugs or betulinum toxin. Hypokinesia, rigidity, and tremor were present in all patients. Bradykinesia of the hands was assessed with a pegboard test, measuring the time (s) required to invert eight pegs. Pegboard performance of patients was compared with that of 46 age matched controls. Results from [123I] IBZM SPECT were compared with 12 other age-matched controls from an earlier study.

A brain dedicated SPECT system, the Strichman Medical Equipment 810X, was used. Two hours after intravenous injection of approximately 185 MBq [123I] IBZM (Cygne BV, Technical University, Eindhoven), tomographic SPECT studies were performed. A maximum of 12 slices was made, starting at the orbitomeatal line and proceeding parallel to it (300 μal; intercremental distance 5 mm). For analysis of specific striatal [123I] IBZM binding, two slices with the highest striatal activity were summed and a template with fixed regions of interest for the striatum and occipital cortex was placed bilaterally on the summed image.

The ratio of the striatal binding to that of the occipital binding quantities specific binding.

The mean ages (table) did not differ among the three groups (t tests). Left and right [123I] IBZM striatal : occipital ratios were significantly lower in patients than in controls (p < 0.05). The degree of asymmetry between [123I] IBZM ratios for the hemispheres in patients or controls (repeated measures multivariate analysis of variance (MANOVA) tests involving side, group, and sex) and group by sex: P > 0.05). The pegboard test revealed that patients with writer's cramp and controls in either hand (t tests), showing that the patients with writer's cramp did not have bradykinesia.

There was no correlation between age or duration of disease and [123I] IBZM ratios (Pearson's correlation coefficients). None of the variables differed between patients with simple and dystonic writer's cramp.

Our results suggest that the striatal dopaminergic system is involved in writer's cramp given that patients with writer's cramp had [123I] IBZM binding different from that of controls. Unfortunately, lack of an accurate measure of the severity of writer's cramp itself prevented us from studying the relation between severity of dystonia and [123I] IBZM.

We did not find a correlation between severity of dystonia and [123I] IBZM ratios. Atal receptors in dystonia, we measured the availability of striatal D2 receptors in patients with writer's cramp using [123I] IBZM SPECT. Unfortunately, lack of an accurate measure of the severity of writer's cramp itself prevented us from studying the relation between severity of dystonia and [123I] IBZM. We did not find a correlation between severity of dystonia and [123I] IBZM ratios and duration of disease or age. This probably means that the decline in striatal D2 receptors is not linearly progressive but remains stable over many years, which accords with our clinical impression. However, because the preclinical [123I] IBZM ratios of the individual patients were not known, it is hazardous to assess rates of decline from a small cross sectional sample.

The results raise some questions. Firstly, there was a bilateral reduction of available striatal D2 receptors, whereas the symptoms were unilateral and there was no asymmetry between the hemispheres. Bilateral abnormalities of the striatum have also been found by others. This bilaterality probably only means that the abnormalities found are related to particular motor dysfunctions which pass undetected if not properly challenged, as shown by the fact that many patients develop writer's cramp on the left side, if they change to writing with the left hand. Accordingly, it is also not uncommon to find involvement of the left, or fingering hand, in musicians playing keyboards, guitars, or other stringed instruments.

A second question is why the reduced availability of D2 receptors on striatal or pallidal membranes is accompanied by parkinsonism in our patients. According to well known models of basal ganglia function, overactivity of the indirect striatopallidal pathway is usually associated with parkinsonism. Because D2 receptor stimulation inhibits the indirect pathway, by contrast with the D1 receptor driven direct pathway, the decreased striatal D2 receptor binding in writer's cramp indicates disinhibition of the indirect pathway which might be expected to be accompanied by parkinsonism. From this viewpoint, we found that the mean [123I] IBZM ratio to be 1.43 (SD 0.16) in patients with definite hypokinetic-rigid dystonia, which accords with our clinical impression. Therefore, our finding is probably better explained by loss of D2 receptors on cholinergic striatonigral and striatopallidal internuncies in dystonia after perinatal anaphylactic injury and with the well known efficacy of anticholinergic therapy in dystonia.

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