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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 postural or visual 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 Previous reports suggest that cerebellar injuries could also cause it 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 and vomiting, and in other episodes vertigo associated with nausea and vomiting and a deviation to the left while walking. When it disappeared, 48 hours later, he complained that he saw objects as if they were tilted to his right by 30° and they should be rotated antevertically—that is, to his left—so as to be perceived as vertical. He had a slight 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. Fungal photographs were not taken, so that ocular torsion could be excluded. 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 without mass effect, in a territory dependent on the posteroinferior cerebellar artery (PICA), not evident with clinical and imaging studies, produced the tilt effects in this case, because the major infratentorial arteries supply 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 hypothetical lesion. Therefore it is not likely that an associated medullary ischemia could cause the tilt effects in our patient. A medullary 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 18F[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 striatohypocortical drive in writer's cramp.1 In view of the

possible involvement of dopaminergic striatal receptors in dystonia, we measured the availability of striatal D2 receptors in patients with writer’s cramp using \([{}^{123}I]\)IBZM SPECT.

Ten consecutive right handed patients (eight male and two female) were classified into two groups: 5 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 botulinum toxin. Hypokinesia, rigidity, and resting tremor were absent 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. Regions of interest were defined on \([{}^{123}I]\)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 approx. 185 MBq \([{}^{123}I]\)IBZM (Cygne BV, Technical University, Eindhoven), tomographic SPECT studies were performed. A maximum of 12 slices was made, starting at the orbitalineline and proceeding parallel to it (300 μl/slice; interslice distance 5 mm). For analysis of specific striatal \([{}^{123}I]\)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 images. The ratio of the striatal binding and that of occipital binding quantities specific binding.

The mean ages (table) did not differ among the three groups (t tests). Left and right \([{}^{123}I]\)IBZM striatal : occipital ratios were significantly lower in patients than in controls (t test). There was no asymmetry between \([{}^{123}I]\)IBZM ratios for the hemispheres in patients or controls (repeated measures multivariate analysis of variance (MANOVA) tests involving side, group, and group by side: P>0.05). The pegboard test showed that patients performed better 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 \([{}^{123}I]\)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 significantly lower \([{}^{123}I]\)IBZM binding than 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 \([{}^{123}I]\)IBZM. We did not find a correlation between \([{}^{123}I]\)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 \([{}^{123}I]\)IBZM ratios of the individual patients were not known, it is hazardous to assess rates of decline in a small cross sectional sample.

The results raise some questions. Firstly, there was bilateral reduction of available striatal D2 receptors, whereas the symptoms were unilateral and there was no asymmetry between the hemispheres. Bilateral abnormal uptake of \([{}^{123}I]\)IBZM has, however, 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. According to Howe et al. 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 should be accompanied by parkinsonism in our patients. 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. This line with the dopamine hypothesis, we can mean \([{}^{123}I]\)IBZM ratio to be 1-43 (SD 0-16) in patients with definite hypokinetic-rigid symptoms due to multiple system atrophy or progressive supranuclear palsy, which is in the same range as the values obtained in most of the previous studies with writer’s cramp (1-27-1-59).

Therefore, our finding is probably better explained by loss of D2 receptors on cholinergic striatal interneurons rather than D2 receptors on striatal spiny output neurons. The fact that \([{}^{123}I]\)IBZM binding was predominately not interneuronal by fibres from the thalamus suggesting a feed forward modulation from thalamus to striatum. A dysfunction of such thalamo-striatal sensorimotor function—caused by increased activity of striatal cholinergic interneurons is sufficient to account for the decreased density of D2 receptors in our patients. Furthermore, striatal cholinergic interneurons are highly represented in the sensorimotor part of the striatum and are predominantly innervated by fibres from the thalamus suggesting a feed forward modulation from thalamus to striatum. A dysfunction of such thalamo-striatal sensorimotor function—caused by increased activity of striatal cholinergic interneurons is sufficient to account for the decreased density of D2 receptors in our patients. Furthermore, striatal cholinergic interneurons are highly represented in the sensorimotor part of the striatum and are predominantly innervated by fibres from the thalamus suggesting a feed forward modulation from thalamus to striatum. A dysfunction of such thalamo-striatal sensorimotor function—caused by increased activity of striatal cholinergic interneurons is sufficient to account for the decreased density of D2 receptors in our patients.