Acute upsidedown reversal of vision in verteobasilar ischaemia

Acute upsidedown reversal of vision is an uncommon and little known phenomenon consisting of transient complete 180 degree inversion of the visual image. The pathogenesis and the anatomical sites of this dysfunction are unknown. Lesions involving cortical areas, mainly the parieto-occipital region, or the vestibulocerebellar system have occasionally been documented.1 2

We observed two patients who experienced this bizarre visual illusion, both revealing features of verteobasilar ischaemia.

Patient 1, a 69 year old woman, was admitted because two weeks earlier she had experienced sudden malaise, sweating, nausea, vomiting, right occipital headache, followed by a 180 degree vertical inversion of the visual image, lasting about 20 minutes. Two similar episodes had occurred the day before admission. On admission the patient was alert, cooperative and well-oriented. The neurological examination was normal. In particular, no neuroophthalmological abnormalities were found on clinical examination. Blood parameters, urine, chest and ECG proved normal. Cervical radiographs revealed osteoarthrosis with osteophytes and narrowing of disc space C6-C7. EEG was normal. Brainstem auditory evoked potentials (BAER) revealed increased latency of V wave on right stimulation. Cerebral CT and MRI (figure) showed an ischaemic-like lesion, 2 cm diameter, in the right cerebellar hemisphere in the territory of the medial branch of the posterior inferior cerebellar artery (PICA), without mass effect. Moderate periventricular white matter abnormalities coexisted. Four vessel cerebral angiography revealed a right vertebral artery stenosis (50%) and two small arteriovenous malformations in the course of the right ascending cervical artery; a decreased flow in the basilar artery was noted. Ticlopidine 250mg daily was given and the patient was discharged. No further attacks or other neurological disturbances occurred during the next two years.

Case 2, a 52 year old woman, with a 40 year history of bilateral chronic otitis with residual deafness, had recent recurrent episodes of sudden swelling, vertigo and occipital headache, dizziness, sometimes followed by a transient loss of consciousness. The whole episode usually lasted about 30–40 minutes. Frequently, at the height of dizziness, the patient complained of a 180 degree vertical visual inversion of images. These episodes occurred monthly. On admission, the neurological examination was normal. Rare, isolated, left-sided jerks of horizontal nystagmus, recorded by ENG. A 10 mmHg difference between right and left brachial arterial pressure (right > left) was noted. Ultrasound vascular investigations (Doppler cortico-vertebral echotomography and cerebral transcranial Doppler) revealed a left subclavian artery stenosis with a steal syndrome. Cerebral SPECT, CT and MRI proved normal. BAER was unavailable due to the peripheral hearing loss caused by chronic otitis. Cerebral angiography was refused. Flunarizine 10mg daily was given and the patient was discharged with a warning to avoid strenuous physical activities, especially those involving upper limbs and neck. No further episodes were reported in the subsequent six months.

These two women presented episodes of vertically inverted vision—upsidedown phenomenon—associated with clinical signs and symptoms of verteobasilar insufficiency. Both reported transient visual inversion of 180 degrees, which was bilateral, of sudden onset and lasting subjective impression of movement (rotatory or horizontal). In the first patient, neuroimaging revealed a right hemispheric cerebellar infarction. In the second, a verteobasilar failure due to a left subclavian stenosis was detected. The pathogenetic mechanism underlying upsidedown visual inversion is unknown. Since the visual images enter the retina inverted, it may be assumed that the upside down phenomenon is the outcome of the mechanisms mediating reversion, even though the anatomical structures involved are unknown. In earlier observations,1–2 parietal and/or occipital lesions were sometimes mentioned as causes of the cortical origin of the dysfunction, probably affecting the integrative control of spatial vision. More recent cases,2–4 documented with neuroimaging techniques, revealed an association with vestibular/cerebellar lesions, that is, verteobasilar TIAs, Wallenberg’s syndrome and also cerebellar infarct in two cases.2 3 In our patients, the relationship between the vertical visual inversion and the signs and symptoms of verteobasilar insufficiency, without evidence of cerebral damage, supports the idea that a transient inactivation of infratentorial structures may cause this unusual phenomenon. Besides the integrity of the visual system, space visual perception needs a flow of extraretinal information, mediated by the vestibular and cerebellar systems.5 6 It has been suggested that damage to such structures may cause tilt and complete inversion of the visual space.7 The upside down phenomenon may occur following dysfunctions at various levels of the vestibular cerebellar-ocular system mediating the stabilisation of the visual function so that cortical involvement is not indispensable.

LETTERS TO THE EDITOR

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Subcortical environmental reduplication: SPECT findings in a patient with a right thalamocapsular haemorrhage

Recently Nighoghossian et al reported the case of a patient with a previous history of a left fronto-basal haemorrhage, who developed environmental reduplication following an infarction of the retrotrenticular portion of the right internal capsule. SPECT revealed right fronto-parietal cortical hypoperfusion. A similar disorganised misidentification syndrome was described previously in a patient with a right thalamic haemorrhage, but its functional correlate using SPECT was not studied.8 We describe the neuroimaging and cognitive functioning of a case of environmental reduplication associated with a right thalamocapsular haemorrhage.

A 71 year old ambidextrous man suddenly developed a left-sided weakness and mild dysarthria. He had had hypertension but no history of previous cerebrovascular events. Neurological examination revealed a dense left hemiplegia, and a left sensory loss affecting all modalities. The left hemiplegia was complete, and there was no evidence of visual or auditory extinction on double simultaneous stimulation. He showed left hemispatial neglect on drawing, and on a letter cancellation task he only crossed targets on the right side of the paper. He did not deny his left hemiplegia, but he had a tendency to attribute it to previous "chest problems". He reported a feeling of nonbeing of his paralysed left arm, and also said that he had three left legs and a strange left arm crossed over his chest. The patient said that he could walk almost normally and repeatedly tried to walk up and down without losing his left hemiplegia. He was alert and oriented to time and person, but not to place. While he

Figure Axial MR T2-weighted image showing a high signal area in the territory of the medial branch of the right PICA.
correlated with the correct name of the hospital, he located in the city. Despite being repeatedly informed that the hospital was in Malaga, he denied it and was convinced that it was a new branch in the city he named. Although he remained in the same room during his admission to hospital, he insisted he had been moved daily to identical hospitals (all with the same name) that were located in neighbouring cities (Torremolinos, Fuengirola, Marbella).

On the day before death, the patient had a detailed neuropsychological examination. He was found to have a WAIS verbal IQ of 94 and a performance IQ of 51. On the Wechsler Memory Scale (WMS), his memory quotient was normal (99 points); his scores on immediate history recall and associated learning were average though his performance on the visual reproduction subtest was below average. There was no evidence of confabulation in response to items of the Mercer’s confabulation battery, but he did poorly on tests thought sensitive to frontal lobe dysfunction (Wisconsin Card Sorting Test: cases B and D; normal range = 4–6). Trail-making test (part A) (below percentile 10). He also showed a severe impairment on visual-perceptual tests (Visual Form Discrimination (14 points; normal range = 23–32); Facial Recognition (15 points; percentile rank = 1). His language was almost intact, except for a mild visual naming impairment (Boston Naming Test, 38 points (maximum = 60)).

CT and MRI scans showed a right thalamic haemorrhage with extension into the posterior limb of the internal capsule, corona radiata and ventricular system. Mild symmetrical periventricular white-matter changes compatible with leukoaraiosis were also observed. Regional cerebral blood flow was studied with "Tc"-HMPAO and SPECT, using an Elscint Apex 609 RG gamma camera. Focal blood flows were analysed semiquantitatively in twelve circular regions of interest which were placed over the cortical mantle in three successive slices. Asymmetry indices (AI) for each lobe were calculated using the following formula: (R-L)/(R+L) × 100. A marked decrease of perfusion was observed in the right thalamus and basal ganglia as well as in the left cerebral cortex. Hypoperfusion was also noted in widespread cortical regions of the right hemisphere affecting mainly the frontal lobe (AI = −29.6), and to a lesser extent the parietal (AI = −10.8) and temporal (AI = −10) lobes (negative AI values indicate left sided hyperactivity relative to the right sided activity).

The assessment of neuropsychological functions in our case of subcortical environ- mental reduplication revealed more pervasive deficits than those observed in the patient reported by Nighoghossian et al., but similar to those of previous cases showing evidence of environmental reduplication and cortical involvement. Moreover, the combination of deficits on nonverbal mem- ory, awareness, visual-perceptual skills and reasoning abilities supports the view that environmental reduplication and cortical involvement are related. The same results are also found in multifactorial delusional misidentification syndrome.

Some previous reports emphasised the association during admission to hospital, the recognition of the bilateral frontal-lobe and right hemisphere cortical involvement, while others suggested that unilateral (right) lesions of either the fronto-parietal or parieto-temporal cortices are sufficient to cause it. In our patient, right thalamocapsular damage may have induced functional depression of various distant but anatomically connected cortical areas. Data from SPECT, however, revealed secondary cortical deactivation affecting mainly the right frontal cortex. In this context, we suggest that functional deactivation of the right cortical mantle, in addition to thalamocap- sular injury, may underlie environmental reduplication and its associated neuropsychological deficits. In addition, given that environmental reduplication probably requires preexisting brain pathology (for example, cortical atrophy) besides the specific sites of brain damage in the right hemi- sphere, the presence of leukoaraiosis in the case by Nighoghossian et al. and in our own patient might be another risk factor for developing it after acute stroke.

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Evidence for presynaptic inhibition on trigeminal primary afferent fibres in humans

In a recent study we have shown that a condi- tioning electrical stimulus applied to the trigeminal afferent fibres of intensity below the reflex threshold (Th) produces an early short-lasting (0–95 ms; 3 times the perceptive Th) of the facial nucleus and the pretibial branch is also reported. There is a clear strict similarity of the curves of the R2 and soleus H reflex inhibition. Identical findings have been observed in two other normal subjects. By exploring the upper curve in fig 1 it is apparent that the long-lasting inhibited by stimulating the hypoglossal nucleus. Afferent discharge of the soleus H reflex is due to the thalamus. It is suggested that the thalamus to spindles causing homonymous Ia inhibition in the soleus motor neurons. This explains why this early facilitation is lacking in the R2 reflex curve (lower curve in fig 1).

The histograms on the right in fig 1 show the development of the R2 and H reflex depressions after acute intravenous adminis- tration of TRH. It has been found that a single subcutaneous injection of high- dose TRH (1-2.5 mg/Kg) produces dra- matic and long-lasting (1–2 hours) increase of the afferent discharge in healthy patients with amyotrophic lateral sclerosis. We have observed a similar but short-last- ing (10–20 minutes) enhancement of the soleus Ia presynaptic inhibition in normal