SHORT REPORT

Right side neglect in right cerebellar lesion

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Abstract

A patient is described who developed right side hemineglect after a right cerebellar lesion. This spatial disorder was interpreted as a secondary effect of a deficit of the motor organisation in the right hemisphere due to left frontal diaschisis. The pathological base may be the interruption of a highly integrated system which includes the lateral cerebellum and the contralateral frontal lobe.

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Historically, the role of the cerebellum has been confined to the domain of motor control. Recent experimental and clinical findings as well as neuroimaging studies have provided parallel evidence that the cerebellum controls not only movement but also sensory function and cognition.

So far, a great deal of neuropsychological evidence supports the notion that the cerebellum modulates linguistic operations, planning, learning, attention, and affective behaviour.^{1,2} A clear demonstration of the role of the cerebellum in processing spatial information was provided by experimental studies adopting the Morris water maze paradigm in mice.³ However, the role of the cerebellum in human spatial behaviour is less clear. Although different degrees of impairment have been documented in various spatial tasks in patients with cerebellar lesions,⁴⁻⁶ only occasionally has a spatial deficit reached clinical importance (see, for example, the case of spatial dysgraphia⁷); this is consistent with the demonstration that the cerebellum participates in the visual and kinaesthetic control of movement.⁸ Recently, the role of the cerebellum in line bisection judgements has been demonstrated by functional MRI in normal subjects.⁹ However, signs of hemi-inattention in patients with cerebellar lesions have never been reported.

We report on a patient who, after a right cerebellar haemorrhage, developed one of the most typical disorders of spatial cognition, spatial hemineglect, which is the inability to pay attention to the side of space contralateral to the cerebral (supratentorial) lesion, in the absence of primary sensory-motor disorders.

Case report

CLINICAL DESCRIPTION

The patient was a right handed, 78 year old woman with 5 years of formal education. History was negative for neurological and psychiatric diseases and the patient was described by her family as able to normally perform everyday living activities. In February 2000 she presented with severe cerebellar symptomatology due to right cerebellar haemorrhage. Brain CT disclosed a large right cerebellar haemorrhage spilling blood into the ventricles, also involving the vermal region. Because of three ventricular hydrocephalus, the patient underwent right ventriculoperitoneal derivation. She was admitted to the rehabilitation unit 3 months after the surgery. On admission her equilibrium remained severely impaired. The patient could not stand without assistance and minor movements of her head and body caused vertigo, nausea, and vomiting. A physical examination showed mild postural tremor and marked hypotonia of the right limbs, mostly the arm. Finger to nose testing elicited marked intention tremor and dysmetria in the upper right arm. Examination of ocular movements showed slower than normal smooth pursuit movements and rightward hypometric saccades; spontaneous nystagmus was occasionally present. The patient complained of diplopia. Oculomotor examination confirmed a deficit of the abducens on the right. Tendon reflexes were normal as well as plantar reflexes, sensory examination, and visual field. Speech was severely reduced and dysarthric but comprehension was good. The patient tended to use the left hand when asked to use objects or in spontaneous movements. However, the possibility of interpreting this tendency as right motor akinesia was questionable because of the motor limitation due to the right cerebellar syndrome. The neuropsychological examination disclosed signs of right hemi-inattention.

Evolution

In subsequent weeks the patient showed progressive improvement of the cerebellar syndrome. At the end of June, when she was discharged from the rehabilitation unit, she was still unable to move without assistance. However, she no longer complained of vertigo and nausea and was more cooperative and oriented; her speech was still dysarthric but initiative and spontaneity were nearly normal. Despite reha-

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Figure 1 (A1 and A2) Brain CT showing the right haemorrhage extending to the vermis and the three ventricular hydrocephalus; (B1 and B2) evolution of the cerebellar haemorrhage after the ventriculoperitoneal derivation.

bilitative therapy, signs of neglect were still present even if the patient tried to apply compensatory strategies. In mid-July 2000 the patient was again studied as an outpatient. Stance and gait were improved but still impaired. Slight right hemi-inattention was still evident. No sign of neglect could be documented at the end of July.

NEUROIMAGING

Computed tomography

A large right haemorrhage extending to the vermis and the coexisting three ventricular hydrocephalus (fig 1 A) was seen. A second examination (fig 1 B) showed the right cerebellar haemorrhage in evolution and resolution of the hydrocephalus after the ventriculoperitoneal derivation.

SPECT

A ^{99m}Tc -ECD SPECT performed 3 months after the onset confirmed an important hypoperfusion in the right cerebellar hemisphere, a marked hypoperfusion in the left frontal lobe, and to a lesser extent in the right frontal lobe. There was no sign of hydrocephalus.

NEUROPSYCHOLOGICAL EXAMINATION

General neuropsychological examination

On admission to the rehabilitation unit in May 2000 the patient was not very cooperative. Psy-

chomotor activities and speech initiation were markedly reduced. Her mood was depressed. Perseverative behaviour was evident both in verbal and non-verbal tasks. Language was markedly reduced, hypofluent, and dysarthric, but lexically and grammatically correct. In the next 3 weeks, the patient improved and a formal examination was possible in short sessions. Naming was pathological because of visual errors; single word and sentence comprehension were preserved. Repetition was normal. In reading tasks the patient produced errors on the right half of the words (errors on the right 18/60; errors on the left 9/60; ambiguous 5/60); writing was severely impaired because of the dysmetric movements of the right hand. Memory (immediate and delayed recall of 15 words) was only mildly impaired (adjusted score: 20 *v* 4.4; cut off: 28.52 *v* 4.69); forward and backward digit and spatial span were pathological (verbal span 3 *v* 2; spatial span 3 *v* 0). Verbal fluency was virtually abolished (no item produced); despite the right cerebellar syndrome, both limb and oral praxis were preserved. Constructional praxis was difficult to evaluate because of the dysmetric movements. Raven's matrices were only mildly reduced (adjusted score: 16.2; cut off 18.96) with perseverative behaviour.

Figure 2 Omission of the stimuli on the right side in the simple cancellation task.

Spatial tasks

The patient was right handed; however, because of the right cerebellar syndrome, she spontaneously tended to use her left hand. Thus, she performed some of the tasks requiring a motor component with the left hand.

Cancellation tasks—On a simple cancellation task (lines scattered on a sheet) performed with the right hand the patient omitted the stimuli on the right half (fig 2) (omissions on the left: 21/31 *v* 6/29; χ^2 13.4; $p=0.003$). A similar tendency to omit the stimuli on the right side was detected when the patient performed the task with her left hand. In subsequent weeks her performance improved and she did not omit stimuli in the line cancellation task; however, she persisted in exploring the left half of the sheet first and oriented her attention to the right half of the sheet only when all the stimuli on the left were cancelled.

Word reading—The patient was given 20 compound nouns to read; she produced 5/20 errors on the left side and 12/20 errors on the right side ($\chi^2=3.88$; $p=0.049$). Errors were mostly visual.

Copying—In drawing a landscape, the patient produced only the first element on the left. When asked to complete two, three dimensional figures she omitted four elements on the right and one element on the left.

Number location—The patient was asked to point to numbers symmetrically distributed on a vertical sheet (10 on the left and 10 on the right). The pointing was performed with the left hand. The patient was accurate in pointing to the numbers located both on the left and on the right, but she needed significantly more time to point to the stimuli located on the right (mean time: left 4.86 s; right 12.25 s; one way analysis of variance (ANOVA): $F=4.618$; $p=0.05$).

Personal neglect—When asked to use an object within her personal space, the patient spontaneously used her left hand but never demonstrated clear asymmetries in spatial exploration of her body. In particular, the patient was able

to correctly use a comb and put on her glasses. **Premotor neglect (directional hypokinesia) component: line bisection (motor and non-motor condition)**—Two conditions were compared¹⁰: in the first condition the patient was asked to mark the midpoint of the line with a pencil held in her right hand (motor condition); in the second, the patient was asked to say when the examiner, who was moving a pen along the line from left to right, had reached the midpoint (non-motor condition). Eight lines of different lengths displayed horizontally and in random order on a sheet of paper were presented in the two distinct conditions; the patient displaced the subjective centre leftward 8/8 in the motor condition and 5/8 in the non-motor condition. Deviation towards the left from the subjective centre (expected in right neglect) was significantly larger in the motor than in the non-motor condition (motor condition: mean 39.00 (SD 28.88) mm; non-motor condition: mean 6.33 (SD 6.88) mm; *t* test 2.68; $p=0.016$).

EXTINCTION PHENOMENA

Single and double stimuli were presented in random order in the visual and sensory modalities. The patient never failed on single stimuli either in the visual or the tactile modality. A non-significant tendency to right side extinction of double stimuli was present in the visual modality (six out of 10 double stimuli extinguished on the right; three out of 10 stimuli extinguished on the left). No extinction was detected in the tactile modality.

Discussion

Hemineglect is a spatial cognition disorder typically characterised by hemispheric lateralisation. Thus, in our patient, the presence of hemineglect suggested a “focal” lesion. In other words, the patient presented something more than the general reduction of performance in visuospatial tasks⁴⁻⁶ reported in other cerebellar patients. Due to the absence of significant lesions in the supratentorial structures, we attributed neglect to the cerebellar damage. We have mentioned that the patient also had hydrocephalus; this could be responsible for some cognitive impairment such as a memory defect, but signs of neglect, and in general “focal signs”, are not typical in this disorder. In addition the hydrocephalus had resolved when the patient was tested.

The hemineglect was associated with the so called cognitive-affective syndrome,¹¹ dominated by the “frontal” signs often described in patients with cerebellar lesions. Both “neglect” and “frontal lobe” syndrome improved over time in our patient, on a parallel with the cerebellar syndrome, reinforcing the hypothesis of a causal relation between the cerebellar damage and the “cognitive” manifestations.

Functional (SPECT) findings were consistent with the “frontal” lobe syndrome and with the side (right) of the neglect. Confirming previous reports on crossed cerebellocerebral diaschisis,¹² they showed a bilateral frontal hypoperfusion, marked in the left frontal lobe, thus congruent with the clinical manifestation

of right side neglect. The finding of a bilateral hemispheric hypoperfusion is also consistent with previous reports suggesting that right side hemispatial neglect is mainly produced by bilateral cerebral lesions that more extensively affect the left hemisphere.^{13 14}

A theoretical account of hemineglect based on the input-output response chain¹⁵ distinguishes between perceptual and premotor (also defined as directional hypokinesia) varieties of neglect for extrapersonal space; the first is closer to perception, the second to motor programming, with segregated neural correlates (parietal *v* frontal).^{16 17}

Considering the role classically assigned to the cerebellum in motor control, the spatial disorder presented by our patient might be interpreted as premotor neglect—that is, a disorder of spatial exploration due to a defective motor programme. Indeed, the patient scored worse in the line bisection task when it required motor execution than in the condition in which no arm movement was required. We have to acknowledge that the patient also showed hemineglect in apparently pure perceptual tasks, such as word reading. However, the rightward hypometric saccades we documented in the clinical examination could have been to some extent an expression of the patient's difficulty in exploration tasks in which the only motor component is represented by eye movement. If this is true, the relatively good performance in non-motor line bisection could confirm that this task makes scarce demands on motor and exploratory resources.¹⁸

It is known that the lateral cerebellum is important for controlling both saccadic and visually guided limb movements.¹⁹ It is also known that a lack of programming of saccades towards contralesional space may be seen in patients with unilateral neglect.^{20–22} Thus, according to the premotor theory of neglect,²³ the patient's spatial disorder could be interpreted as a secondary effect of a deficit in motor organisation involving both limb and eye movements. The pathological basis of this deficit would be found in the interruption of a highly integrated functional system which

includes the lateral cerebellum and the contralateral frontal lobe.

Our patient confirms the role of the cerebellum in “high level” spatial functions. Further evidence is necessary to specify the nature of cerebellar control in the exploration of the hemispace.

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