

LETTERS TO THE EDITOR

Alexia without agraphia or hemianopia in parietal infarction

Alexia without agraphia (pure alexia) may result from damage to the pathways conveying visual input from both hemispheres to the dominant angular gyrus, which itself remains intact but disconnected from the visual regions.¹ The most common lesion involves the left occipital lobe, and may also compromise the splenium of the corpus callosum (occipital or splenio-occipital alexia).^{2,4} Less often, pure alexia results from a lesion either in the occipitotemporal paraventricular white matter,^{3,4} or more superiorly and rostrally in the parieto-occipital or parietal white matter (subangular or paraventricular alexia). To date, only three cases of pure alexia without visual field defect have been described in patients with proven parieto-occipital lesions.³⁻⁷ The responsible lesion was surgical in one case⁵ and intracerebral haematoma in two cases.^{6,7} We now report a patient with alexia without agraphia or visual field defect, in whom MRI demonstrated a subcortical infarction in the left parieto-occipital area.

A right handed normotensive physician had mitral valve prolapse, paroxysmal atrial tachycardia, and intermittent atrial fibrillation. He was 66 years old in 1982, when he suffered a right frontal lobe infarction. Since the diagnosis of a cystic glioma was also entertained, he had had open craniotomy, which confirmed the presence of an infarct. CT scans, carried out at the time of infarction and yearly thereafter, showed a large, non-enhancing, low density cystic lesion in the right frontal region. His neurological examination was normal aside from the cognitive deficits described below. Extensive psychometric testing in September 1985 demonstrated mild attentional, visuospatial and constructional deficits, and mild impairment in his ability to learn new nonverbal material.

Confrontation naming was normal.

On the evening of 21 January 1986, he felt very anxious, sensed that something was wrong, and went to bed. The following morning, on awakening, he was unable to read the newspaper or the phone book. He had no difficulty with verbal output. His wife noticed that he was mildly confused, with difficulties in verbal comprehension. On examination four hours later he was oriented to person, place and time. The quantity and grammatical content of verbal speech production were normal, and there were no paraphasic errors. Repetition of "no ifs, ands, or buts" and "Methodist Episcopal" was normal. Comprehension for conversation and three part commands was normal. He named visually presented common objects but had mild difficulty naming small parts of objects. He could name saturated colours, match colours, and point to a named colour. Reading was severely impaired. He had some impairment naming letters and arabic numerals and understanding some isolated words. He could not understand the meaning of sentences, including simple written commands. Compared with his severe alexia, he had only minimal agraphia when copying written material and writing to dictation. For example, when asked to write "Today is a sunny day in Southern California" he wrote "Totay is a sussy day in Southern Califortia." Although he could not perform written calculations because of difficulty reading Arabic numbers, he performed mental calculations well. The remainder of his mental status and general neurological examination was unchanged from his baseline. In particular, confrontation visual fields were normal.

Over the course of the following 48 hours, the patient's reading ability improved significantly. He could read and understand simple sentences, although not complex ones. Writing became entirely normal. Reading comprehension then gradually improved in the ensuing weeks, and extensive psychometric testing in April 1986 showed similar deficits to those observed in September 1985. At this time, reading comprehension was normal, although the patient stated that his reading speed was slower than before, and he had lost interest in reading.

MRI (1.5 Tesla) brain scan (fig) on the second day of alexia showed: 1) the pre-

existing right frontal lesion; 2) an area of increased signal intensity in the paraventricular white matter of the left parieto-occipital junction, underlying the junction of the left angular gyrus and left lateral occipital gyrus (fig A); and 3) an area of increased signal intensity in the overlying parieto-occipital cortex (fig B). MRI brain scan carried out four months later again showed the left subangular lesion but no longer showed the cortical high intensity signal. This lesion was seen on T_2 weighted but not on T_1 weighted images. CT brain scan, carried out on the second day of alexia, showed the pre-existing right frontal lesion but failed to demonstrate the left parieto-occipital lesion that was observed with MRI. Six weeks later a CT brain scan demonstrated the new left subangular infarction. Octopus perimetry done five months after the stroke was normal.

The patient fulfilled the criteria for the diagnosis of pure alexia established by Benson and Geschwind.² He had severe disturbance of reading comprehension, relatively preserved writing, and absence of aphasia or dementia. The pre-existing deficits secondary to the frontal lobe lesion could be separated from his more recently developed reading deficit. The impairment of written calculations, a frequent finding in pure alexia,⁸ was attributed to difficulties in comprehension of written numbers, since mental calculation abilities were spared. No elements of the Gerstmann syndrome were present.

MRI demonstrated an area of increased signal intensity (low T_2) in the white matter immediately underneath the cortex at the left parieto-occipital junction. The lesion was observed on T_2 weighted images on the second day, when T_1 weighted images and CT were negative, a pattern of abnormality that is characteristically seen in acute cerebral infarction.⁹ The CT demonstrated the lesion six weeks later. The transient increased MRI signal intensity in the overlying parieto-occipital cortex was felt to reflect reactive hyperaemia associated with the acute infarction.

To our knowledge, this is the first report of pure alexia without hemianopia due to a parieto-occipital ischaemic infarction. The lesion was confined to the paraventricular white matter subjacent to the left angular gyrus and spared the optic radiations. Three previously reported patients had pure alexia without hemianopia due to lesions with similar anatomical localisation.⁵⁻⁷ Nonetheless, localisation was less precise because the lesions were acute surgical or intracerebral haematomas presumably with associated mass effect.

We would like to express our appreciation to Ms Barbara Reader for her assistance in the preparation of this manuscript.

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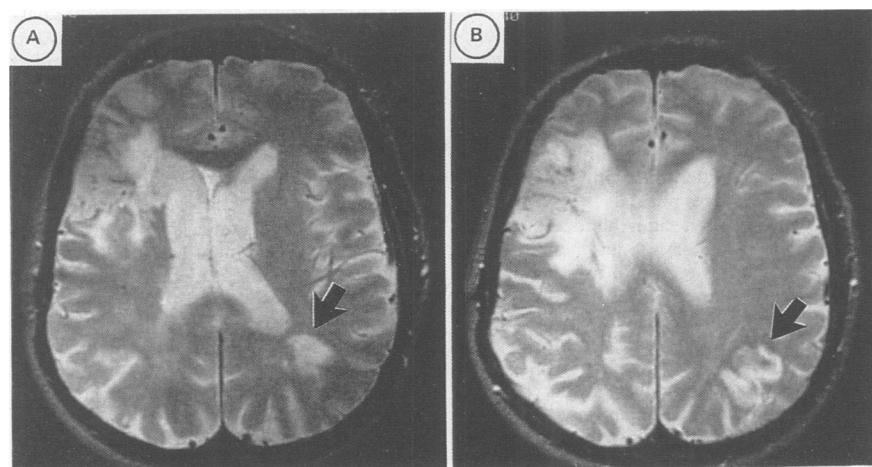


Figure MRI brain scan of pure alexia carried out on day 2. Spin echo, T_2 -weighted images ($TR = 2000$ ms, $TE = 70$ ms). Note subcortical area of increased signal intensity underlying the left parieto-occipital junction A) and area of increased signal intensity in the overlying parieto-occipital cortex B).

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Minor hemisphere syndrome following left hemispheric lesion in a right handed patient

Some degree of unawareness of hemiplegia occurs in about one third of cases of right hemisphere patients with left hemiplegia.¹⁻⁴ Anosognosia for hemiplegia may be associated with left hemiasomatognosia. Such patients behave as if the left half of their body was no longer part of themselves.⁵ In contrast to the relative frequency of such phenomena following damage to the right hemisphere, anosognosia and hemiasomatognosia have rarely been reported following left hemisphere lesions. None of the reported cases has concerned authentic right-handed patients and the degree of language impairment has often been unknown.

We describe a case of a strongly right handed patient with asomatognosia and anosognosia for right hemiplegia. She exhibited other deficits relating to the so-called minor hemisphere syndrome, and had no language disorders.

On the thirteenth day after aortic valve replacement for aortic regurgitation, the patient, a 68 year old right handed woman, developed an acute hemiplegia. On the fourth day following onset, neurological examination showed massive right motor deficit affecting the face, the arm and the leg. Right plantar reflex was extensor, there was severe hypesthesia and tactile extinction on the right side of the body, and right homonymous hemianopia on confrontation. Language and praxis were normal on bedside evaluation. The patient had motor impersistence and strongly tended to keep her head and eyes turned to the left, even on verbal stimulation from the right. CT scan at 10 days post onset revealed a left hemispheric infarct involving the territory of the middle cerebral artery, both deep and superficial, and the territory of the anterior choroidal artery.

The following observations were gathered during the first three weeks following the stroke, during which period the neurological condition of the patient remained essentially unchanged. Right sided visual neglect was seen on the dot cancellation test: she failed to cancel 7/9 dots in the right half of the test sheet, although she did not miss any of the eight dots in the left half. The patient was

presented with a list of 40 pairs of items (10 pairs of digit names, 20 pairs of object names, 10 pairs of sentence fragments). She showed clearcut right auditory neglect: she correctly reported all of the 40 items presented to her left ear, but none of those presented to her right ear. She was largely unaware of her hemiplegia. When asked if she could move her right hand, sew, or knit, she answered that she could. When asked if she could move her right leg, she answered positively and moved her left leg as a proof. Sometimes, the patient admitted that she needed some help with walking or running. Once, when asked if she could cut her meat and eat all by herself, she affirmed that she could, if only helped to sit in her bed. She occasionally showed some abstract knowledge of her deficit. She said that doctors had told her she had hemiplegia, but that she did not believe it, since she was not paralysed and could walk.

The patient also showed a variable degree of right asomatognosia. In several instances, when shown her right hand, she would answer that it was the hand of a corpse that had been introduced into her bed. However, she sometimes correctly identified the hand as her own. Touching it with her left hand apparently facilitated the identification. When asked to designate her right hand she was initially unable to find it in the bed. Twelve days after onset, she could correctly designate her right arm, leg, eye and cheek.

Her spontaneous speech was considerably aposodic and she was initially unable to sing at all, although she previously used to participate in an amateur choir. Two weeks after the stroke, her few attempts at singing were very much out of tune. Moreover, her perception of melodies was also impaired. Five days post-onset, she could not identify common tunes that were hummed to her, but recognised them readily as soon as the lyrics were added. Three weeks later, she could identify five out of 10 popular tunes whose melodies were sung to her.

The patient described herself as completely right handed, and denied having been forced to use her right hand as a child. There was no record of any significant cerebral trauma in birth or early childhood. She was submitted to a 24 item questionnaire about her preferred hand or foot in various everyday-life activities. She always unambiguously chose the right side. Her parents, grand-parents, and two children were all reported to be right handed.

Language evaluation was normal in all respects. Spontaneous speech, repetition, object and picture naming, designation, fluency in controlled association, comprehension, and reading were flawless. She was too awkward with her left hand to allow evaluation of writing. There was no left hand apraxia. Visual identification of objects, colours and faces was normal.

The patient was a right handed woman, with no family history of left handedness. After a left parieto-temporal infarction, she showed several symptoms that usually follow right hemispheric lesions, that is, symptoms encompassed in the so-called minor hemisphere syndrome. She had right hemispatial neglect,⁵ right hemiasomatognosia, anosognosia for right hemiplegia, motor aposodia⁶ and a severe impairment in identifying and producing musical tunes⁷ but no aphasia.

Two main conclusions can be drawn concerning the patient's pattern of cerebral dominance. First, her right hemisphere was

certainly dominant for language. If the left hemisphere was dominant for language, or if language was bilaterally represented, the extensive left hemispheric softening would have caused aphasic symptoms, which was not the case. Second, her left hemisphere was strongly dominant for manuality, spatial attention, and body schema.

Few cases of right asomatognosia or anosognosia for right hemiplegia have been reported. Dronkers and Knight⁸ have described a left handed patient with clear-cut anosognosia for right hemiplegia, right spatial neglect and aposodia, following left hemispheric infarct. Cambier et al⁹ have reported a similar case, but their patient seems to be ambidextrous. Such is also the case of the patient reported by Hermann and Pötzl.¹⁰ Among their patients with left-hemispheric lesions, Hécaen and Sauguet¹¹ mention right hemiasomatognosia in two out of 47 left-handed patients, but in none of 293 that were right-handed. Gross and Kaltenbäck¹ have described two patients with such a denial of right hemiplegia, including one case with hemiasomatognosia. These patients were both aphasic, but there is no mention of their manual dominance. Cutting² has reported three instances of denial of hemiplegia out of 22 testable right hemiplegic patients. Their manual dominance and degree of language impairment, however, are not reported.

The dominant hemisphere for body schema is usually the right hemisphere. However, as we have just seen, considerable evidence exists of reversed dominance, as revealed by right asomatognosia and anosognosia for right hemiplegia. The left hemisphere is usually dominant for manuality and language, although reversed dominance in this domain is also well known. But are all the different patterns of co-dominance concerning language, manuality and body-schema biologically possible? Or are some of these functions necessarily supported by the same hemisphere (or by opposite hemispheres)? We shall consider each pair of functions in turn. First, the left hemisphere is usually dominant for both language and manuality. Our patient falls into the 4% of those that are right handed with right hemispheric dominance for language, according to Rasmussen and Milner's figures¹² (for discussion, see Geschwind and Galaburda¹³). Second, manuality and body schema are usually dissociated since in most right handed subjects, the right hemisphere is dominant for body schema. The patient reported by Dronkers and Knight⁸ shows exactly the opposite pattern of dominance. However, Hécaen and Sauguet¹¹ mention several left handed patients with left asomatognosia, suggesting that the right hemisphere can occasionally support both functions. The pattern of impairment in our patient demonstrates that that can also occur for the left hemisphere. Third, language and body-schema are usually dissociated, but one hemisphere can occasionally be dominant for both functions, as illustrated by the patients of Gross and Kaltenbäck,¹ who have at the same time aphasia and anosognosia for hemiplegia following left hemispheric lesion. Our patient's dominance pattern is the exact opposite of the pattern which prevails in most of those that are right handed, since her left hemisphere is dominant for body schema, and her right hemisphere for language.

The case favours the idea that there is no obligatory link between any two of the three cerebral functions we have considered, and