ALEXIA WITHOUT AGRAPHIA OR HEMIANOPIA IN PARITIAL 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. The most common lesion involves the left occipital lobe, and may also compromise the splenium of the corpus callosum (occipital or splenio-occipital alexia). Less often, pure alexia results from a lesion either in the occipitotemporal paraventricular white matter, 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. The responsible lesion was surgical in one case and intracerebral haematoma in two cases. 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 defects, 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 paraplectic 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 "Today is a sunny day in Southern California." Although he could not perform written calculations because of difficulty reading Arabic numerals, 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 A) 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 (fig A) and T1-weighted images of pure alexia. 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. 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 separable from his more recently developed reading deficit. The impairment of written calculation abilities was severe.

MRI demonstrated an area of increased signal intensity (low T2) in the left occipital lobe, without mass effect. The lesion was observed on T1-weighted images on the second day, when T2-weighted images and CT were negative, a pattern of abnormality that is characteristic of infarction. 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 parieto-occipital white matter subjacent to the left angular gyrus and spared the optic radiations. The lesion had been successfully treated with anticoagulation, and was considered to be unrelated to the previous infarction. The patient has continued to make good functional progress, and no neurological deficit was noted at the time of writing.

Figure MRI brain scan of pure alexia carried out on day 2. Spin echo, T2-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 unawarness of hemiplegia occurs in about one third of cases of right hemisphere patients with left hemiplegia.\textsuperscript{1,4} Anosognosia for hemiplegia may be associated with left hemisomaotognosia. Such patients behave as if the left half of their body was no longer part of themselves.\textsuperscript{1} In contrast to the relative frequency of such phenomena following damage to the right hemisphere, anosognosia and hemisomaotognosia have rarely been reported following left hemispheric 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 somatognosia 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 left 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 was put in 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 clearrcut right auditory neglect: she correctly reported all of the 40 words 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 felt any weakness, numbness, or sensory loss in her right hand, arm, or leg, she answered negatively. 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 somatognosia. 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 aprosodic 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 perinatal or early childhood brain trauma in her life. 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 hand for primary or secondary tasks (cooking, playing with the grand- and great-grandchildren, and two children) and was therefore considered 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. She showed 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. There was no right hemispatial neglect,' right hemispatial anosognosia, anosognosia for right hemiplegia, motor aprosodia and a severe impairment in identifying and producing musical tunes but no aphasia.

'Main two conclusions is now 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 aphasia, according to her, which was not the case. Second, her left hemisphere was strongly dominant for manuarity, spatial attention, and body schema.'