Alexia without agraphia in multiple sclerosis

Abnormalities of the afferent visual system, especially the optic nerve, are common in multiple sclerosis. They are the initial symptoms in about one third of the patients and occur in almost all of the patients during the progression of the disease. On the other hand, retrochiasmatic lesions of visual pathways rarely cause clinical manifestations of multiple sclerosis. An incidence of 1-3% of homonymous defects has been reported in several series and there is only one patient reported with alexia and agraphia.1 We report a patient with alexia without agraphia that was the initial manifestation of the disease.

A 26 year old right handed man was admitted with a week long inability to read and diminished vision in the right visual field. He was oriented and tried to be cooperative. Affect was depressed. Speech was fluent and comprehension and repetition were normal. He was unable to read but he could write fluently on dictation or spontaneously. He was able to name objects, letters, colours, and numbers but was unable to read the whole word. He had no visual disorientation and memory deficits. His visual acuity was 20/30 OD and 20/30 OS. He had a dense right inferior quadrantanopsia to 1/4 and complete homonymous hemianopia to I/4 on the Goldmann perimeter test. There was a minimal left hemiparesis. His blood cell counts, urinalysis, blood chemistry, ECG, and chest radiograph were normal. Antinuclear antibody, antcardiolipin antibody, anti-DNA, and C3 and C4 titres were negative. Examination of CSF showed normal pressure, no cells, and normal protein and glucose concentrations. Microbiological and serological studies of CSF gave negative results. Immunoelectrophoresis of CSF showed oligoclonal bands and raised IgG concentrations. Visual evoked responses were abnormal, with prolongation of P100 latencies to 140 ms in the right and 160 ms in the left eye. Brainstem auditory evoked responses were abnormal on the right and somatosensory evoked responses showed evidence of central delay from the left tibial nerve. Brain MRI showed lesions that were hypointense on T1 weighted series, and hyperintense on T2 weighted series in the posterolateral part of the left post, posterior horn of both internal capsules, left occipital cortex, the splenium of the corpus callosum, and subcortical and paraventricular white matter (figure). The lesions were thought to be compatible with multiple sclerosis. According to the Posner criteria, he was evaluated as having laboratory supported definite multiple sclerosis (LSDMS B).2 He was treated with intravenous methylprednisolone (1 g daily for five days) and then this treatment was replaced by 64 mg oral methylprednisolone daily which was gradually reduced. His condition improved and he was able to read at the time of discharge. At the follow up, three months after discharge, he was feeling well and his neurological examination was normal.

Although symptomatic lesions of the optic nerves and chiasm are common in multiple sclerosis, symptomatic retrochiasmal disease and higher level visual disturbances are unusual.1 Most lesions in tempo-occipital white matter are found to be clinically silent. Our patient presented with alexia without agraphia and MRI disclosed lesions in the left occipital lobe, paraventricular white matter, and the splenium of the corpus callosum. Associated features included right homonymous hemianopia and minimal left hemiparesis. Lesions involving the medial portion of the left occipital lobe that spare the angular gyrus cause alexia without agraphia. Alexia without agraphia was first described by Dejerine in 1892 as a result of lesions disconnecting the pathways between the occipital lobes and the temporo-parietal association areas. Later Geschwind postulated that any lesions preventing bilateral visual stimuli from reaching the left angular gyrus might produce this syndrome. Although it is not necessary to involve the splenium, the lesion is usually demonstrated with involvement of the splenium of the corpus callosum.3 Our patient had the criteria for the diagnosis of alexia of Benson and Geschwind—namely, severe disturbance of reading comprehension, linguistically correct writing, normal oral spelling, and absence of aphasia and dementia. According to the classification introduced by Damasio and Damasio, our patient could be type II; found patients with right hemianopia without colour anomia or verbal anomia. In this type, there is damage to optic radiations or to the calcarine region, and compromise of the interhemispheric pathways within the left occipital lobe, due to a paraventricular lesion. It has been postulated that the absence of colour anomia or verbal anomia may be due to the fact that both left lingual and parahippocampal cortices, and left hippocampal formation are involved, which may be true for our patient. In addition, the preserved ability to name letters may be due to an intact anterior part of the corpus callosum or may be the result of the residual ability of the right hemisphere to read as Geschwind suggested.1

Alexia without agraphia is most commonly caused by ischaemic infarction due to occlusions of the posterior cerebral artery.4 It has also been reported in patients with spontaneous intracerebral haemorrhage, arteriovenous malformations, intracranial neoplasm, brain abscess, and migraine.5

Alexia without agraphia as the initial manifestation of multiple sclerosis is unusual and this is the first case reported to result from multiple sclerosis.

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Urinary retention in bilateral pontine tumour: evidence for a pontine micturition centre in humans

Normal micturition is the result of a coordinated action of the muscles of the bladder and urethral sphincter. A complete transection of the spinal cord above the sacral level usually leads to a lack of coordination between bladder and urethra (detrusor-sphincter dyssynergia) and urinary retention whereas a suprapontine lesion does not lead to dysynergia but to incontinence.6 The fact that detrusor-sphincter dyssynergia was found only in patients with well defined neurological lesions of the spinal cord suggests that coordinated voiding is regulated by neurological centres above the spinal cord.7 Barrington showed in the cat that lesions in the dorsal part of the pontine tegumentum produce inability to empty the bladder.8 He suggested the presence of a so-called pontine micturition centre and that detrusor-sphincter coordination takes place in the pons.9 In neurological textbooks the pontine micturition centre is described as a pontine pathology among the possible causes of urinary retention in humans, and