LETTERS TO THE EDITOR

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%–3.5% 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 V/4 and complete homonymous hemianopsia 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, anticardiolipin antibody, antiDNA, 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 pro-

longation 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 pons, 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 Poser criteria, he was evaluated as having laboratory supported definite multiple sclerosis (LSDMS B3).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 retrochiasmatic disease and higher level visual disturbances are unusual.¹ Most lesions in temporo-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 hemianopsia 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 Déjerine in 1892 as a result of lesions disconnecting the pathways between the occipital lobes and the temporoparietal 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 writ-

ing, 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 in patients with right hemianopsia without colour anomia or verbal amnesia. 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 amnesia may be due to the fact that both left lingual and parahippocampal cortices, and left hippocampal region are intact, which may be true for our patient.⁴ 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.³

Alexia without agraphia is most commonly caused by ischaemic infarction due to occlusive cerebrovascular disease involving the posterior cerebral artery.⁵ 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 lack of coordination between bladder and urethra (detrusor-sphincter dyssynergia) and urinary retention whereas a suprapontine lesion does not lead to dyssynergia but to incontinence.¹ 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.¹ Barrington showed in the cat that lesions in the dorsal part of the pontine tegmentum produce inability to empty the bladder.² He suggested the presence of a so called pontine micturition centre and that detrusor sphincter coordination takes place in the pons.² In neurological textbooks there is no mention of a pontine pathology among the possible causes of urinary retention in humans, and

Axial T2W MR images showing hyperintense lesions involving (A) subcortical, periventricular white matter, left occipital cortex and (B) splenium of corpus callosum.

also in research literature the reports are scant.³⁻⁵ We describe a patient presenting with paraparesis and urinary retention in whom a pontine tumour extending bilaterally was discovered.

A 69 year old woman with hypertension and type II diabetes had complained of weakness in the lower limbs with a fluctuating but progressive course since November 1994. On February 1995 she had an acute deterioration and could not walk or urinate. At admission there was pronounced weakness (MRC 2) in the lower limbs (worse on the right) and slight weakness (MRC 4) of the proximal and distal muscles of the upper limbs. Tendon reflexes were brisk in the upper limbs and hypoactive in the lower limbs. Plantar response was extensor bilaterally. Sensory examination was normal. There was bladder distension at percussion and 1200 ml urine were evacuated by catheterisation. The course was characterised by spontaneous improvement of strength in the lower limbs and reappearance of spontaneous, although incomplete, voiding. After two weeks there was worsening of lower limb weakness, definitive urinary retention, the appearance of left internuclear ophthalmoplegia, and mild dysphagia.

Lumbar puncture showed normal pressure values. Examination of CSF was normal. Urodynamic studies showed a hyporeflexia of detrusor muscle without detrusor-sphincter dyssynergia. Motor conduction velocities and compound muscle potential amplitudes, sensory conduction velocities and sensory nerve potential amplitudes, and late response latencies were normal. Tests of autonomic nervous system function were normal. Echography excluded a pelvic tumour. A complete spinal MRI study was normal. Brain MRI showed a lesion in the pons which was hypointense in T1 weighted images, and non-homogeneously hyperintense in T2. Gadolinium thiopentate infusion profoundly enhanced the lesion, which was bilateral, extended from the pontine base to the tegmentum, and slightly compressed the floor of the fourth ventricle (figure). Some small lesions, which did not show enhancement, were present in the hemispheric periventricular white matter and were ascribed to vascular leucoencephalopathy. The clinical and imaging findings suggested a low grade glioma. Because of family reasons the patient was transferred to a hospital in another country and died after five months.

The patient we report had urinary reten-

tion and a pontine tumour extending bilaterally and involving the pontine tegmentum. Other possible causes of urinary retention were excluded. The pontine micturition centre responsible for urine emptying is localised in the cat in the medial part of the dorsolateral pontine tegmentum.² Physiological studies in experimental animals show that the centre is represented in both sides of the pons and that bilateral lesions are necessary to cause severe micturition disturbances. The occurrence of urinary retention in patients with pontine disease has been rarely reported. Ueki in 1960 and Reiner and Gabreels in 1980 mentioned urinary retention in some patients with pontine tumours.^{3,4} These reports, lacking modern neuroimaging, did not correlate the site and the extension of the lesions with the urinary disturbances. Up to now only one other case with MRI showing a bilateral pontine lesion (thought to be a developmental abnormality) and voiding dysfunction has been reported.⁵ It is noteworthy that urinary retention is not mentioned in circulatory disorders involving the pontine tegmentum. The patient we describe provides further clinical evidence for the importance of the pons in the pathophysiology of human micturition and suggests some points:

(1) In humans, as in experimental animals, the pontine micturition centre seems to be located in the dorsal tegmentum confirming that normal micturition is a brain-stem reflex and not a simple sacral one,

(2) In humans, as in experimental animals, a bilateral lesion in the pontine micturition centre seems to be necessary to cause voiding dysfunction indicating that the integrity of only one pontine micturition centre is sufficient to maintain normal voiding,

(3) The necessity of a bilateral lesion accounts for the rarity of clinical reports and for the fact that urinary retention has been reported essentially in tumours and not in pontine vascular lesions.

A large series of patients correlating site and extension of the lesion demonstrated by MRI with the urodynamic findings would be necessary to ascertain the role of the pons in human voiding disorders and confirm our data. In the meantime, from the practical point of view, neurologists should be aware that urinary retention can be due to bilateral pontine disease involving the tegmentum.

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Chronic inflammatory demyelinating polyneuropathy with multifocal CNS demyelination in an Afrid

The occurrence of a syndrome that combines chronic inflammatory demyelinating polyneuropathy (CIDP) with multifocal CNS demyelination is now well recognised but rare.¹⁻³ The nature of the CNS demyelination is still uncertain. Although the clinical manifestations and MRI findings resemble multiple sclerosis, no pathological studies are yet available. All patients reported so far have been ethnically European. We have recently encountered a case in an Afrid. In view of the rarity of multiple sclerosis in Afrid populations it seems of interest that the patient should be documented.

The patient was an ethnically Afrid Sudanese girl aged 21 years with no European antecedents. Her parents were normal but were remote cousins. When aged between 1 and 2 years she had developed a right lateral rectus muscle weakness which had recovered over the course of some weeks. When aged 9 years she had developed weakness of her right hand so that she was no longer able to hold a pen to write. A cranial CT performed at the time was normal. This disability recovered fully after two to three weeks. Her recent history was of numbness in her right arm, trunk, and leg for the previous two months with weakness of her right arm and leg and poor balance on her feet. She had experienced burning paraesthesiae distally in all four limbs and "crawling" paraesthesiae in her right upper limb.

Both optic discs were pale. Her visual acuity was normal and her visual fields were full. Bilateral palmar-mental reflexes were present. She showed clumsiness for fine finger movements on the right, mild weakness in her right hand, and ataxia on tandem walking. Apart from sluggish brachioradialis and knee jerks, her tendon reflexes were absent, as were her abdominal reflexes. Both plantar responses were extensor. The appreciation of light touch and pinprick was impaired over her right face, arm, trunk, and leg and joint position sense was mildly impaired in the fingers and toes bilaterally. Other sensory modalities were normal. Her peripheral nerves were not thickened. Examination of other systems was negative.

Routine haematological, biochemical, and autoantibody screening was negative, including testing for anti-G_{M1} ganglioside antibodies; her serum proteins and white cell enzymes were normal. Examination of CSF disclosed oligoclonal IgG bands but was

Brain MRI: T1 weighted images after gadolinium thiopentate infusion. Left: coronal section showing abnormal enhancement in the pons more pronounced on the left. Right: sagittal section showing that the lesion extended from the base to the pontine tegmentum.