Upsala J Med Sci 106: 205–210, 2001

Aphasia in Multiple Sclerosis

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ABSTRACT

Aphasia has rarely been reported in multiple sclerosis (MS). In this article a case with MS presenting as purely global aphasia during an exacerbation of MS is reported with clinical and MRI (magnetic resonance imaging) findings. MRI demonstrated giant plaques mimicking a cerebral tumour with surroundings characterised by oedematous transformations in the left frontal and parietal lobes.

INTRODUCTION

Aphasia as a clinical manifestation of cerebral MS is a rarity [7, 8]. The frequency of aphasia in MS was estimated in several series to range from 0.7% to 1.0% [1]. The presence of aphasia has even been regarded as incompatible with the diagnosis [7].

In this article, a patient with MS, in whom neurologic examination showed no signs other than global aphasia in her third attack, is presented with MRI findings.

CASE REPORT

A 22 year-old, right-handed woman was admitted to a university hospital experiencing weakness of her left arm and leg as well as urinary incontinence for fifteen days prior to her admittance.

Neurologic examination revealed mild to moderate weakness of the left arm and leg. Deep tendon reflexes were brisk on the left. She also had extensor plantar response on the left. Cerebrospinal fluid examination (CSF) and evoked responses were normal. MRI demonstrated demyelinating plaques in frontal and occipitoparietal regions and posterior limb of internal capsule in periventricular white matter on right cerebral hemisphere, and in white matter adjacent to the occipital horn on the left cerebral hemisphere. The surroundings of active plaques were oedematous (Fig. 1). On history, six months before her admittance, the patient had visual difficulty and urinary incontinence, which had improved within a month, and therefore the patient was considered to suffer from second exacerbation of MS and was diagnosed clinically as probable MS according to Poser's criteria (CPMS A1). The patient was treated with oral metylprednisolon. Clinical findings nearly improved within a couple of weeks.

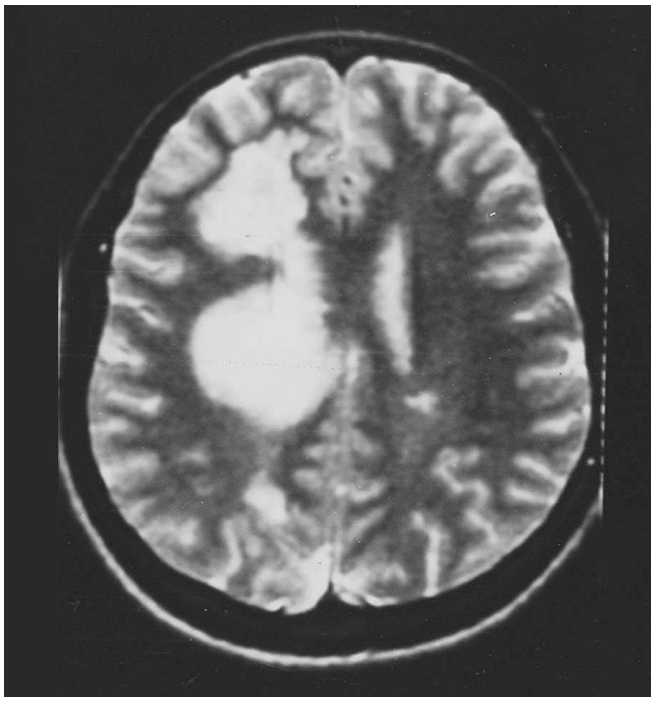

Fig. 1. Axial MRI scan (T_2-weighted sequence): One year before the evolution of aphasia, in the second attack, large hyperintense plaques simulating a tumoral lesion in the patient's right.

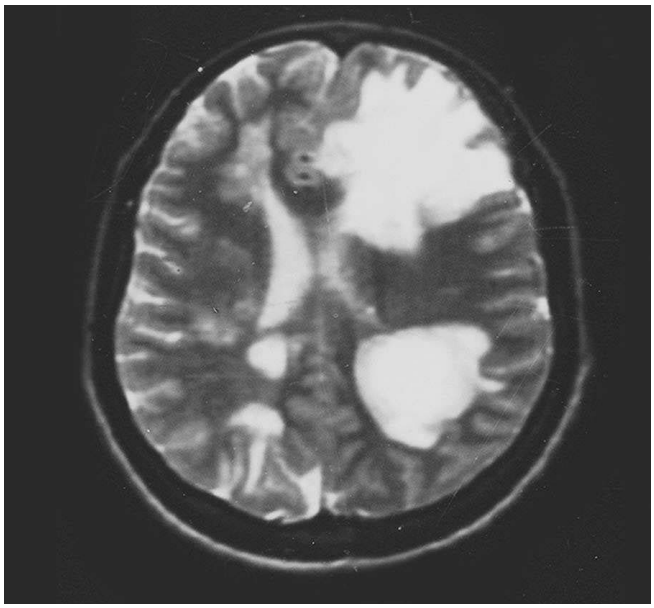

Fig. 2. Axial MRI scan (T_2-weighted sequence): During the aphasia, in the third exacerbation, new giant white matter lesions with mass effect especially in the patient's left cerebral hemisphere. The lesions on the right hemisphere in the previous attack have disappeared, and new giant plaques having shift effect on the lateral ventricle have occurred on the left side.

The patient was readmitted to the hospital because of speech impairment after one year. On neurologic examination there were no other findings except global aphasia. Brainstem auditory evoked potentials (BAEPs) and somatosensory evoked potentials (SEPs) from median and tibial nerve stimulation performed bilaterally. BAEPs had revealed prolongation in the interpeak latencies I–V and III–V on the right side. SEPs from left median nerve stimulation showed absence of cortical components. MRI showed numerous hyperintense lesions bilaterally whose surroundings were highly oedematous in left frontal and parietal regions in T2-weighted images. These lesions had shift effect on the left lateral ventricle (Fig. 2). In this MRI, right cerebral lesions found in the previous MRI had disappeared, whereas there were new lesions not seen in the previous attack in the left cerebral hemisphere. According to Poser's criteria, the patient was accepted clinically definite MS (CDMS A1). Global aphasia gradually improved with oral metylprednisolone treatment. She had only slight difficulty to repeat long sentences.

In her fourth attack, fourteen months after global aphasia, neurologic examination revealed mild right hemiparesis. Deep tendon reflexes were very brisk on the right and enhanced on the left. Plantar responses were extensor bilaterally. Vibration and joint-position senses were impaired in the right upper and lower limbs. CSF examination disclosed increased immunoglobulin G index. Oligoclonal bands were present in the CSF. Evoked potentials were performed bilaterally. Visual evoked responses

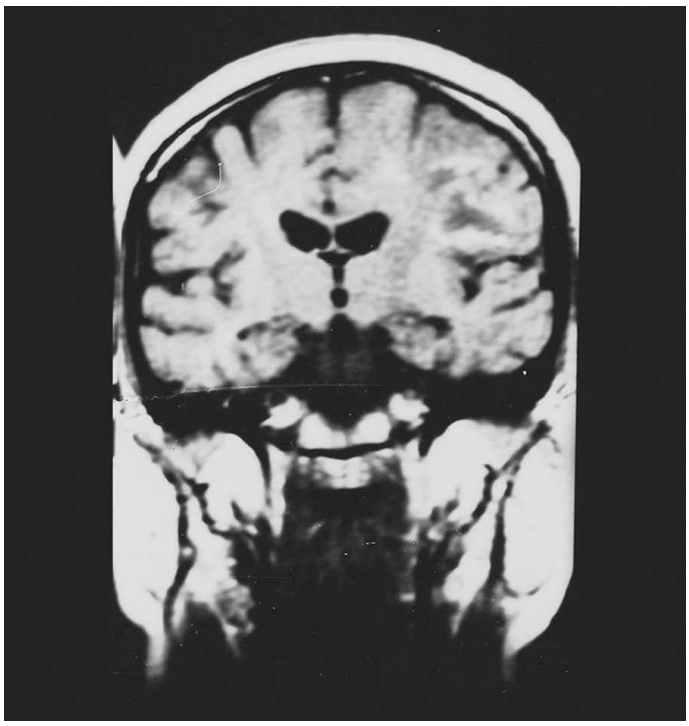

Fig. 3.

to pattern shift stimulation were abnormal bilaterally. SEPs from right tibial nerve stimulation showed absence of cortical components. BAEPs revealed a delay in the latency of the fourth peak on the left and loss of fourth and fifth waves bilaterally. MRI demonstrated multifocal demyelinating plaques showing linear irregular contrast enhancement in both cerebral hemispheres (Fig. 3). The patient was treated with intravenous metylprednisolone (1 g/d) for five consecutive days. The neurological findings nearly improved.

DISCUSSION

First, Gussenbauer reported a patient with aphasia, suspected of having a left hemispheric tumour, in whom autopsy disclosed only multiple sclerosis plaques in 1902 [1]. Since the first description, there have been only a few additional anecdotal reports on the occurrence of aphasia as part of MS exacerbation [1, 4, 8].

Achiron et al. [1] reported two MS patients with acute onset of motor aphasia. Serial MRIs demonstrated the acute appearance of large plaques that are compatible with white matter plaques typical of MS in the left frontal region in one case and the left centrum semiovale in the other patient [1, 2]. In both cases, the anatomic localization of the lesions could account for motor aphasia. It was proposed that only white matter plaques that were sufficiently large to simultaneously involve and disrupt commissural, association and projection fibbers in the dominant frontotemporal region could cause motor aphasia [1].

Although it has been generally accepted that the syndrome of Broca's aphasia is classically caused by lesions within the posterior two-thirds of the third frontal cortical convolutions [1, 8], it has been reported also in subcortical lesions involving the operculum, insula, and adjacent white matter [1]. The concept of diaschisis, whereby impaired function occurs in one area of the brain secondary to an acute focal lesion in a more distant part of the brain, has been cited as one explanation for subcortical lesions causing aphasia [3]. Since demyelinating lesions frequently affect subcortical region [1], and the involvement of these regions by plaques is frequently shown at post-mortem examination [8], it seems puzzling why aphasia is so uncommon in MS [1]. One possible explanation is that MS plaques are generally neither large enough nor dense enough to cause aphasia [4]. It is also possible that function may be maintained by adjacent undamaged fibbers, by supplementary areas of the cortex, or by virtue of an intrinsic functional resistance of the affected areas themselves [8].

Most of the cases with aphasia reported in literature are the patients with motor aphasia [1, 2, 3, 8]. Arnett et al. [2] reported an MS patient with conduction aphasia associated with a large white matter lesion underlying the left supramarginal gyrus [2].

To our knowledge, our patient is the first case of global aphasia in whom MRI findings may be correlated with the clinical presentation during an exacerbation of MS. The first case with global aphasia suffering MS was reported in 1983. The diagnosis of MS had been based on only clinical criteria [4]. In our patient, the plaques seen in MRI during the third attack were large enough to involve all pathways that are responsible for language function. Moreover these giant plaques have been pre-

sented as a cerebral tumour with mass effect and peripheral contrast enhancement. Analysis of MRI findings in patients with MS in several series shows that such giant plaques are uncommon [1].

It has been reported that five patients with MS presenting aphasia have giant plaques on the MRI scan [1, 3]. Therefore the reason of aphasia being a rarity in the MS literature may be the fact that patients have unusually large white matter lesions extending close to the cortical surface [2]. MS plaques typically do not produce mass effect [6]. If areas of demyelination and associated inflammatory cellular response are marked and confluent, this may result in a focal space-occupying lesion often associated with contrast enhancement due to loss of integrity of the blood-brain barrier [5, 9]. This can cause a high signal area on the MRI scan simulating a tumour or an abscess.

In her third attack, our patient's MRI showed large lesions whose surroundings were characterised by the oedematous changes in the left frontal and parietal lobes. Fourteen months after this attack, in her fourth attack, some of these lesions simulating tumoral disease were not seen on MRI scan. Because these multifocal lesions have different shapes, and no mass effect and perifocal oedema, they are compatible with demyelinating plaques of multiple sclerosis. In addition, clinically the patient's speech impairment had recovered. Clinically these relapsing-remitting course and MRI findings showing contrast enhancement led us to consider an exacerbation of MS.

The other point of notice is that our patient's clinical findings were not so severe as expected. Thompson et al. [10] could not find any relation between the degree of clinical disability and the extent of abnormality shown by MRI: patients with clinically benign disease often had extensive abnormalities and those with primary progressive disease had surprisingly few lesions [10].

In conclusion acute aphasia may present as an exacerbation of the disease in MS [3]. The involvement of the cortical white matter junctions including all the anatomic structures, which are responsible for language function, may have caused the global aphasia. With the appearance of giant plaques, MS rarely may be present as a focal cerebral mass, mimicking a glioma [6]. Although MRI findings in MS are not always correlated to clinical findings, and are not predictive of the prognosis, it is import to differentiate the demyelineating lesions of MS from the other conditions.

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