Hemiparetic multiple sclerosis

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Abstract

Eight patients are described who presented with hemiparesis which involved the face in seven. Six of the eight subsequently developed clinically definite multiple sclerosis and in the remaining two patients multiple sclerosis was the likely diagnosis. Magnetic resonance imaging gave useful information about the site of the lesions responsible for the presenting syndrome and provided additional information in support of a diagnosis of multiple sclerosis.

Hemiparesis which involves the face is uncommon as the presenting feature of multiple sclerosis (MS), and may lead to diagnostic confusion, particularly when the syndrome is progressive or stuttering in onset. In recent years various investigations have enabled the diagnosis of MS to be pursued in patients who, because of their unusual clinical picture, previously might not have been suspected of suffering from the disease. Of these investigative techniques magnetic resonance imaging (MRI) of the brain and spinal cord is the most sensitive. In this article patients with hemiparetic MS are described in whom MRI scanning gives useful information about the probable sites of the lesions responsible for the hemiparetic episodes. Other patients with an acute hemiparetic syndrome are described in whom MS is the most likely diagnosis.

Patients

Case 1 In 1975, when 39 years old, this right handed woman experienced an episode of tingling affecting the whole of the left side of her body and over the next three years she had five further episodes all precipitated by exertion. The symptoms lasted for up to three weeks and were accompanied by mild weakness on the left side. Between episodes she was free of either symptoms or signs. At the age of 45 years she had another similar episode of sensory disturbance and was admitted to hospital where within a few hours she developed a left hemiparesis involving the face and limbs. Computerised tomography (CT) of the head was normal. She recovered to near normal over a period of six months. She then started to have intermittent headaches and over the following eight months she had an episode of weakness of the right leg followed by an episode of left hemiparesis. A repeat CT scan showed an area of low attenuation in the right cerebral hemisphere, thought to represent a mature infarct. Routine blood tests, erythrocyte sedimentation rate (ESR), serology for syphilis and autoantibodies were normal or negative. Further investigations were unremarkable including echocardiogram, arch aortogram, carotid ultrasound scan and visual evoked potentials (VEPs).

In 1984 (aged 48 years) there was another episode of hemiparesis involving the right face and limbs which was associated with dysphasia. She was admitted to the National Hospital at which time the weakness had largely resolved and the physical signs were limited to ataxia of all limbs. A further CT scan showed two areas of low attenuation, the first in the white matter adjacent to the right trigone and the second in the left centrum semiovale. MRI at that time showed widespread periventricular lesions and other lesions within the cerebral white matter with lesions bilaterally in the corona radiata immediately superior to the internal capsule (fig 1). The cerebrospinal fluid (CSF) was acellular but showed oligoclonal banding of the immunoglobulins. VEPs and somatosensory evoked potentials (SEPs) were normal.

Case 2 This right handed man developed a mild left hemiparesis, involving the face, when aged 34 years. This resolved completely over a period of two weeks. He remained well until the age of 51 when he became impotent. At the age of 56 years he had an episode of unsteadiness of gait with veering to the right. This lasted a week but did not resolve completely and was followed by a progressive unsteadiness of gait and dragging of the right leg, and later he developed slurring of his speech and clumsiness of the hands. He was admitted to the National Hospital in 1985 when aged 59 years. On examination he had a slurring dysarthria and there were pyramidal signs in all limbs with extensor plantar responses and ataxia of limbs and trunk. Routine blood tests including ESR and serology for syphilis were normal; the anti-nuclear antibody (ANF) was not sought. VEP latency was increased bilaterally and SEPs and brain stem auditory evoked potentials (AEPs) were both abnormal. The CSF was acellular with negative syphilis serology but showed oligoclonal banding. Cranial CT was normal. MRI in 1985 showed several lesions in the white matter of both cerebral hemispheres and in the right internal capsule (fig 2).

Case 3 This left handed man had two attacks of right sided weakness between the ages of 13 and 15 years. When sixteen years

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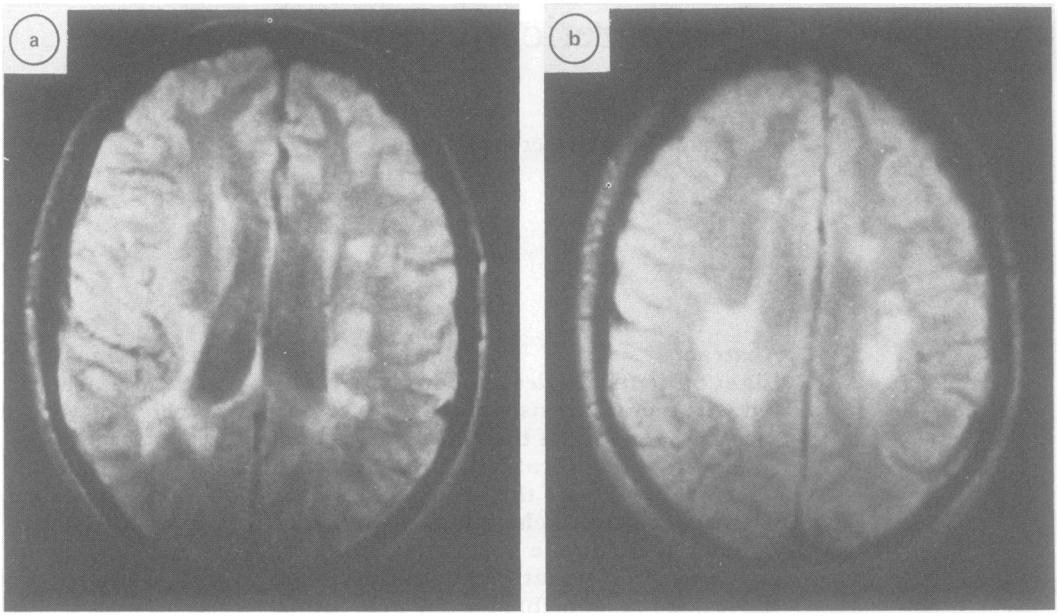

Figure 1 MRI scans at two levels (a, b) in patient 1 (SE 2000/60). There are periventricular lesions and also discrete white matter lesions. The lesions involve the white matter immediately above the internal capsules.

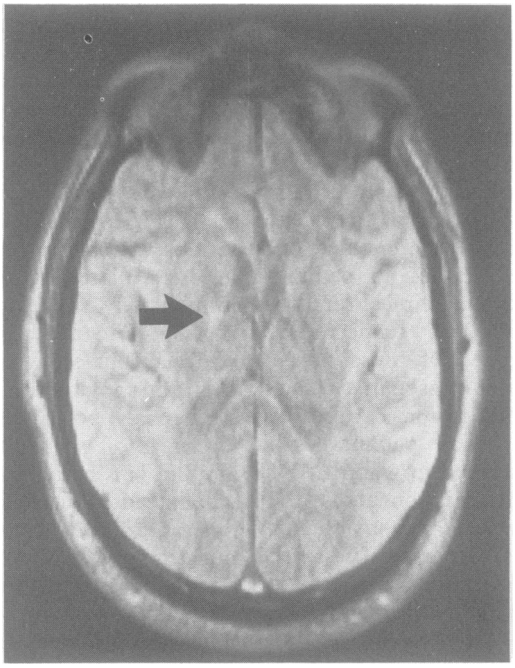

Figure 2 MRI scan in patient 2 (SE 2000/40). There is a lesion in the right internal capsule (arrowed).

old he developed, over a period of 24 hours, a right hemiparesis involving the face which was associated with a right hemisensory impairment, headache and intermittent diplopia. Two weeks later he was admitted to the National Hospital. On examination he had a slurring dysarthria. He had incomplete abduction of the left eye and some nystagmus on lateral gaze to right or left. He had a severe right hemiparesis with an extensor plantar response on the right. The CSF had a normal protein content with six white cells/mm^3. A left carotid angiogram was normal. He recovered within two weeks. At the age of 22 years he had a left hemiparesis and hemisensory disturbance which recovered over eight weeks. A few months later he had an episode of right sided weakness with bilateral visual impairment and intermittent diplopia. This came on within days of a bicycle accident in which he had experienced an apparently minor head injury. He also noticed tingling down the spine and legs on neck flexion. On examination he had some restriction of abduction of both eyes, nystagmus on upgaze and first degree horizontal nystagmus in both directions. There was sensory loss involving all three divisions of the right trigeminal nerve, and both taste and hearing were impaired on the right. In the limbs there was weakness of the right leg with right sided hyperreflexia. The CSF contained six lymphocytes/mm^3 and 0·5 g/l of protein. Bilateral vertebral and right carotid arteriograms were normal.

When he was 46 years he had an episode of tinnitus with right hemisensory loss and diplopia on looking to the right. These symptoms lasted for two weeks. Two years later he developed a right sided weakness over a few hours and three days later he became dysphasic. He was admitted to the National Hospital and found to have a nominal dysphasia and a dense right hemiparesis involving the face with right hemisensory loss. The CSF contained five lymphocytes/mm^3 and a protein of 0·69 g/l and oligoclonal bands were present; the syphilis serology was negative. Routine blood tests were normal including the ESR and autoantibody screen. The CT showed mild cerebral atrophy, cerebellar atrophy and an area of low attenuation in the left corona radiata. The VEP was delayed on the right.

MRI scanning was performed in 1985 (aged 52 years) which revealed lesions in the deep cerebral white matter of both hemispheres and in the left cerebral peduncle. There were also lesions in the posterior limbs of both internal capsules (fig 3). Examination at this time revealed a right hemiparesis and ataxia in both arms. The plantar responses were flexor.

Case 4 In 1979, aged 23 years, this man suddenly developed a sensation of pins and needles down his right side followed by a dense right hemiparesis. These symptoms passed off within minutes but he had nine

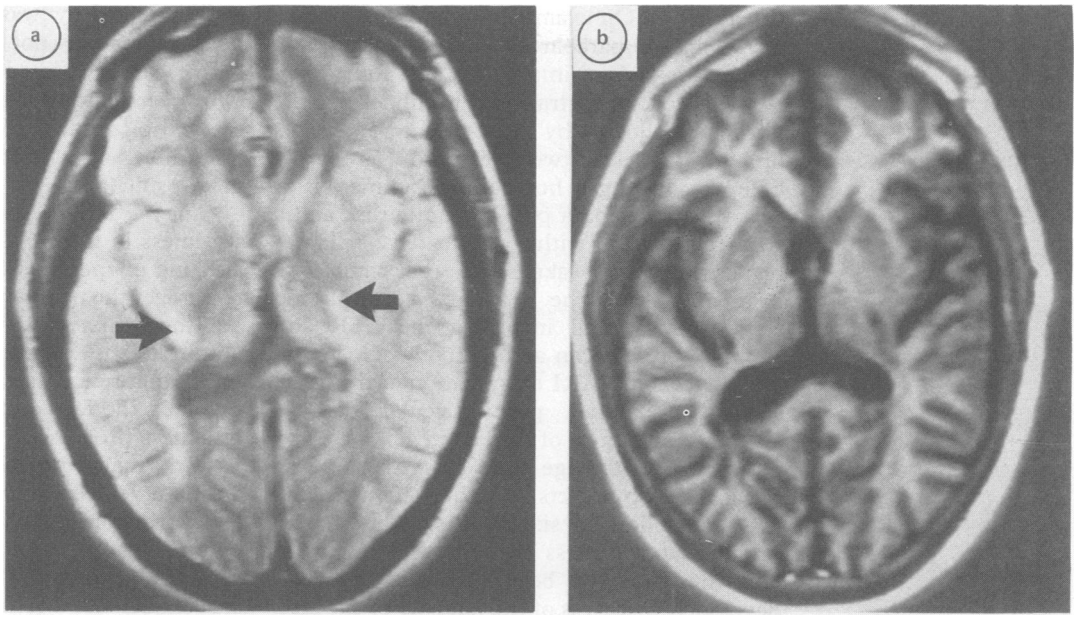

Figure 3 MRI scans in patient 3. The SE 2000/40 (a) image shows lesions in both internal capsules (arrowed) confirmed on the IR 2000/500 image (b).

similar episodes over the next 24 hours each lasting about 10 minutes. He was transferred to the National Hospital the following day and had another three similar episodes affecting the right side the longest of which lasted for two hours and during which examination revealed a right hemiparesis involving the face with hyperreflexia, a right extensor plantar response, and right hemisensory disturbance. He was left with a mild residual weakness which cleared over the next two months. Routine blood tests were normal or negative including ESR, ANF and serology for syphilis. A CT scan showed an area of low attenuation in the region of the internal capsule bilaterally. Four vessel cerebral angiography was normal. He subsequently developed impaired visual acuity bilaterally with right optic atrophy and bilaterally delayed VEPs. Oligoclonal bands were found in the CSF. Cranial MRI in 1985 showed periventricular changes around the margins of the lateral ventricles and discrete lesions within the cerebral white matter. In particular there were lesions in the posterior limb of the internal capsule on the right and there was a lesion involving the posterior aspect of the left lentiform nucleus and pallidum which involved the posterior limb of the internal capsule on the left (fig 4).

Case 5 At the age of 33 years this man developed numbness and weakness in the right leg, followed some months later by similar symptoms in the right hand. He was admitted to hospital the following year where he was found to have first degree nystagmus to the right and a mild right hemiparesis, involving the leg more than the arm, and sparing the face. Myelography revealed no significant abnormality and a CT head scan was normal. Examination of the CSF and the VEPs were normal.

When he was 38 years of age he complained of nocturnal jerking of the right and of intermittent constipation. He noticed that his legs were stiff and this was exacerbated by

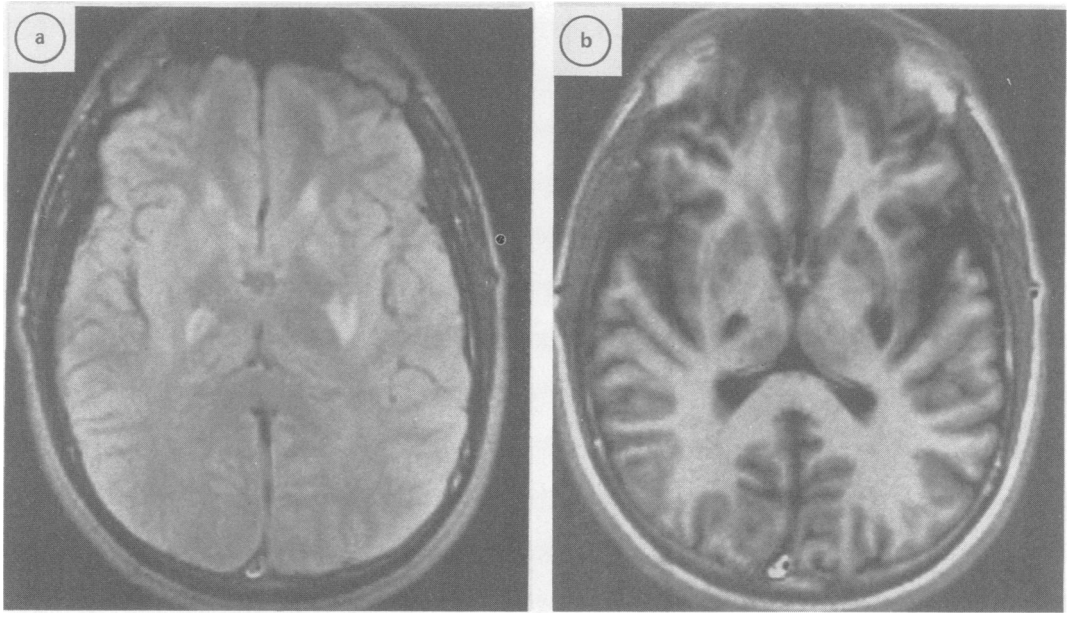

Figure 4 MRI scans in patient 4. The SE 2000/60 (a) image demonstrates lesions in both internal capsules. The anatomical position of the lesions is shown more accurately on the IR 2000/500 (b).

constipation. On examination he was found to have a spastic paraparesis. The following year he complained of impotence and four years later he developed transient numbness of the left hand followed by intermittent pain in his limbs which lasted two weeks and was exacerbated by having a hot bath. Examination at that time revealed a pale left optic disc and a right hemiparesis with a right extensor plantar response. The weakness of the right side progressed over the next five years. Blood tests were normal including the ESR and syphilis serology; an autoantibody screen was not performed. MRI examination at that time showed bilateral periventricular lesions around the margins of the lateral ventricles.

Case 6 At the age of 32 years this woman began to drop objects from her left hand and over the following six months she noticed a progressive weakness of the left hand. A year later her left leg began to drag and she developed weakness of the left side of the face. She was admitted to hospital for investigation in 1984 and at that time the CT head scan and the VEPs were normal. The CSF had a normal cell count with negative serology for syphilis but contained oligoclonal bands. No definite diagnosis was reached. Following her discharge she had several episodes, each lasting a few hours, of blurred vision affecting the left eye. She also noticed a temporary increase in her left sided weakness following a hot bath. She was re-admitted to hospital for myelography which showed a swelling in the cervical spinal cord, and the weakness improved a little following a course of ACTH injections. Later that year there was a progressive decline in the power of her left side and her speech became slurred. She was admitted to the National Hospital for further evaluation. On examination there was bilateral optic atrophy and a left hemiparesis with a left extensor plantar response. The tone in the right arm was increased and all limb reflexes were brisk. The ESR was normal and the syphilis serology was negative; an autoantibody screen was not performed. MRI of the brain was performed during this admission, two years after the first symptom. This showed bilateral periventricular lesions and a lesion in the posterior limb of the right internal capsule (fig 5).

Case 7 This 26 year old woman awoke with weakness in the right arm and leg and a mild slurring of speech. There was some improvement over the next few hours but the following day the weakness was more marked. She was admitted to the National Hospital. On examination there was a right hemiparesis which involved the face and over the next week the weakness progressed. The routine blood tests were normal or negative including ESR, autoantibody screen and the syphilis serology. Evoked potentials, CT head scan and left carotid angiography were normal. The CSF protein and cell count were normal but the IgG was oligoclonal. MRI examination of the brain was performed during her admission and this showed a lesion in the left internal capsule and there were two other lesions seen in the cerebral white matter (fig 6).

Case 8 This 33 year old woman developed sudden blurring of vision in the right visual field of each eye followed by complete loss of vision in the same fields. This was associated with some pain, mainly over the forehead. Five days later she developed a worsening of the headache and some unsteadiness of gait. She was admitted to the National Hospital. The visual acuities were 6/9 bilaterally and there was a right homonymous hemianopia with a mild right hemiparesis. Routine blood tests were normal or negative and these included the ESR and ANF. The CSF contained eight lymphocytes/mm^3 with a protein of 0·55 g/l and oligoclonal bands but negative syphilis serology. A CT head scan was normal but MRI at that time revealed a lesion in the posterior limb of the right internal capsule and periventricular lesions around the margins of the

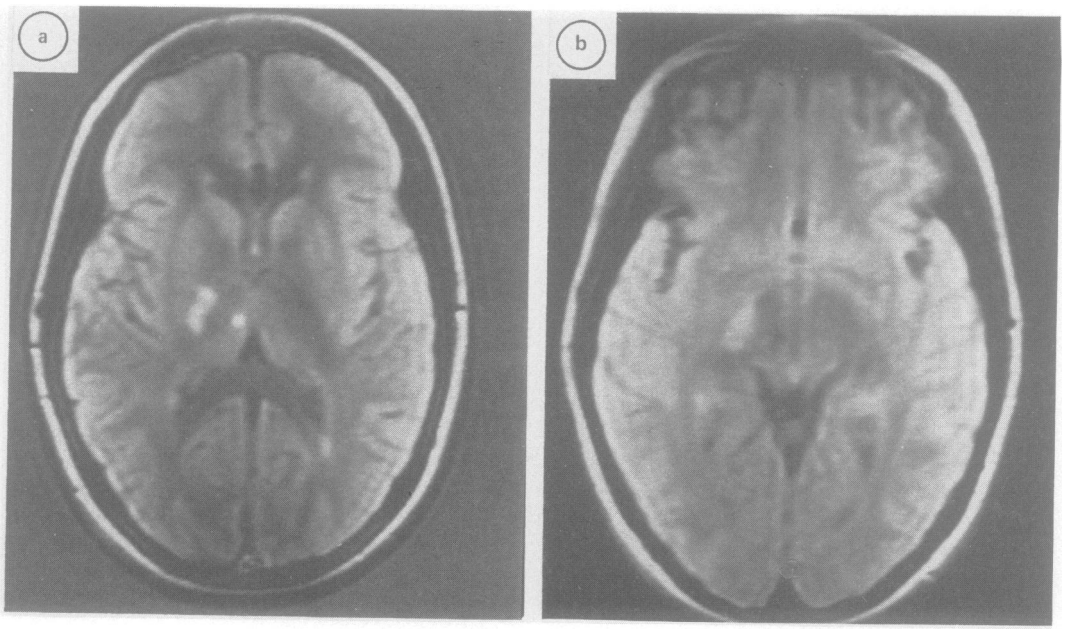

Figure 5 MRI scans (SE 2000/40) in patient 6. There is a lesion in the right internal capsule (a) which also involves the cerebral peduncle (b).

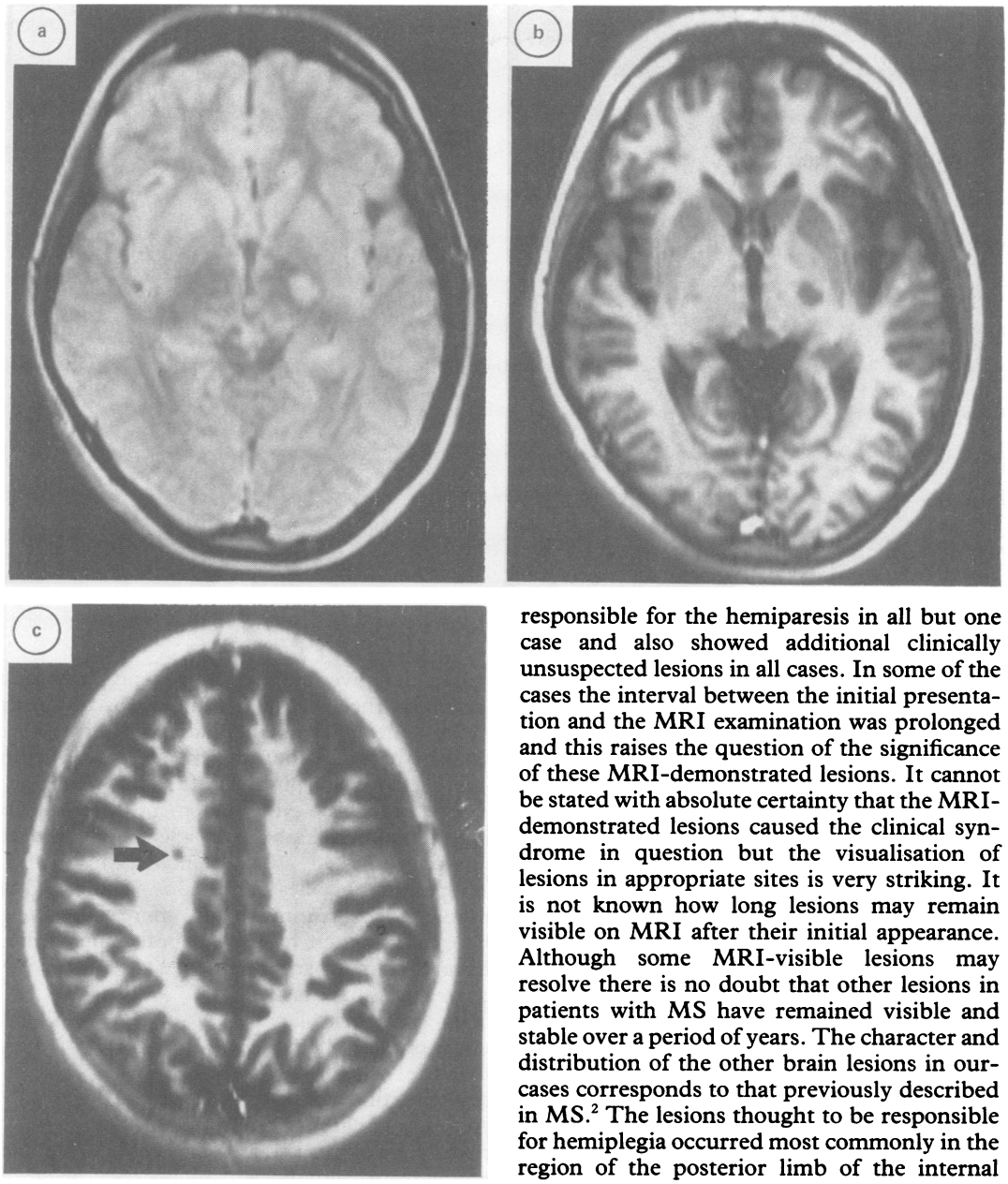

Figure 6 MRI scans in patient 7. There is a lesion in the left internal capsule seen on SE 2000/40 (a) and IR 2000/500 (b) images. There are also other lesions (arrowed) within the cerebral white matter (c) seen on the IR 2000/500 image.

lateral ventricles of both hemispheres (fig 7).

Discussion

The veracity of the diagnosis of MS in the above cases must be considered. Since none of the patients has died post mortem information is not available. The most recent criteria for the diagnosis and categorisation of MS include CSF analysis for oligoclonal bands, evoked potentials and information obtained from imaging techniques.[1] Using these criteria cases[1-6] can be classified as clinically definite MS. Although it is most likely that cases 7 and 8 presented with a first attack of MS they cannot be classified as MS at this time despite the presence of oligoclonal bands in both. In each of these latter cases there was only one clinical attack with appropriate physical signs but the paraclinical evidence of additional lesions (MRI) was obtained during the initial episode without adequate temporal separation.

The MRI scans showed the lesion probably responsible for the hemiparesis in all but one case and also showed additional clinically unsuspected lesions in all cases. In some of the cases the interval between the initial presentation and the MRI examination was prolonged and this raises the question of the significance of these MRI-demonstrated lesions. It cannot be stated with absolute certainty that the MRI-demonstrated lesions caused the clinical syndrome in question but the visualisation of lesions in appropriate sites is very striking. It is not known how long lesions may remain visible on MRI after their initial appearance. Although some MRI-visible lesions may resolve there is no doubt that other lesions in patients with MS have remained visible and stable over a period of years. The character and distribution of the other brain lesions in our cases corresponds to that previously described in MS.[2] The lesions thought to be responsible for hemiplegia occurred most commonly in the region of the posterior limb of the internal capsule, a site containing many large, myelinated fibres, through which runs the pyramidal tract.[3]

In case 5, the only patient in whom the face was spared, the hemiparetic syndrome might be explained clinically by a spinal cord or brain stem lesion; not all patients with clinical evidence of a brain stem or spinal cord syndrome have lesions demonstrable by MRI.[4,5] The dysphasia observed in cases 1 and 3 is not easily explained on the basis of the lesions shown; there did not appear to be appropriately sited cortical or subcortical lesions in these patients. Dysphasia has been described previously in patiens with MS but it is uncommon and there has been discussion concerning the sites of the lesions responsible.[6,7]

It is unclear why hemiplegia involving the face is relatively uncommon in MS especially when the corticospinal tracts are heavily myelinated and have a long course. In a large MRI study of patients with clinically definite MS, lesions within the internal capsule were seen in 11% of patients.[2] To be clinically apparent a lesion must disrupt the function of

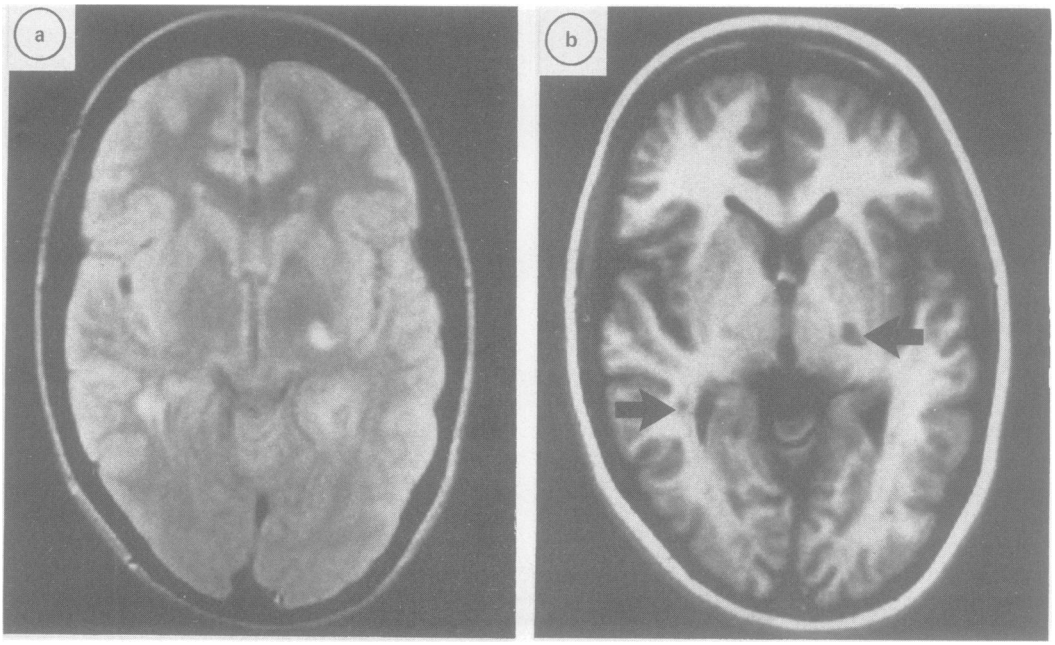

Figure 7 MRI scans in patient 8. There are lesions in the left internal capsule and around the right trigone (arrows). The anatomical site of the lesions is seen more clearly on the IR 2000/40 image (b).

an adequate number of axons in a given pathway and this may explain in part the apparent disparity. Lesions affecting the pyramidal tracts at a more caudal level are probably more common, particularly in the cervical spinal cord,[8] and such lesions are frequently symmetrical in the cord giving a tetraparetic rather than a hemiparetic presentation.

The nature of the hemiparetic episodes is also interesting. Symptoms have sometimes lasted for only a few minutes while others have lasted for months or have been permanent. Some episodes of marked weakness have lasted for several days and then have completely remitted, only to return later on both the same and the opposite sides of the body. Why a patient should have a recurrence of such a rare symptom as hemiparesis, particularly with bilateral non-synchronous involvement, is unknown. Transient episodes, especially those only lasting an hour or two are typical of MS, as is the worsening of symptoms after a hot bath.[9] This may merely be physiological highlighting of an already existing lesion but hemiparesis lasting longer presumably indicates the occurrence of a new lesion.

Hemiparetic MS has been discussed previously but only rarely as a isolated topic. McAlpine noted hemiplegia as an unusual first symptom in MS and commented that these episodes may be brief and recurrent.[10] He also stated that the sudden onset of hemiparesis in MS may cause confusion with a haemorrhagic or thrombotic event;[11] angiography was often performed in our patients and a diagnosis of migraine suggested in several. Kelly[12] notes that the onset of hemiplegia in MS is most commonly subacute and Matthews[13] comments that acute hemiparesis in MS is rare and also appears more commonly in men.

When a hemiparetic episode occurs as a first neurological manifestation a vascular event, tumour, vasculitis, syphilis and encephalitis are among the diagnoses considered and investigations are performed to ascertain the cause and guide the clinician on appropriate management. MRI may be useful in these patients in identifying the causative lesion and also demonstrating other clinically unsuspected lesions. These findings may obviate the need for more invasive investigations such as cerebral angiography.

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