SHORT REPORT

Recurrent myelitis

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Abstract

Three patients presented with acute complete transverse myelopathy which relapsed several times at the same site. These patients, two women and one man, had two to five attacks spanning three to seven years. All patients underwent detailed investigations including a complete myelogram and serial evoked potential studies. Oligoclonal bands were present in the CSF in one patient. Brain MRI was normal in two patients; MRI of the spinal cord was abnormal and showed cord oedema with multiple areas of hyperintense signals on T2 and proton density weighted scans and hypointense signals on T1 weighted images in areas corresponding to the clinical level, suggesting an inflammatory/demyelinating disorder. These patients may represent a relapsing demyelinating disorder restricted to the spinal cord, distinct from multiple sclerosis.

Keywords: complete transverse myelopathy; relapsing; recurrent disseminated encephalomyelitis

Transverse myelitis is most often the cause of acute transverse myelopathy once vascular occlusive disorders and toxic and radiation injuries are excluded. Acute transverse myelitis may be best described as a postinfectious autoimmune demyelinating disorder similar to acute disseminated encephalomyelitis.

Recurrence of isolated myelopathy poses a diagnostic dilemma. Systemic lupus erythematosi,

1 antiphospholipid antibody syndrome,

2 isolated angitis of the CNS,3 HIV,4 herpes simplex infections,5 and spinal vascular malformations6 have been reported to produce recurrent isolated cord syndromes. Recurring spinal cord dysfunction in multiple sclerosis manifests most often as a partial cord syndrome, generally of subacute or chronic onset and with associated evidence of dissemination at onset or in the early follow up period.7

Recurrence of isolated complete spinal cord syndrome presumed to be of demyelinating aetiology has been reported recently.8-11 In some reports there has been substantial evidence on serial MRI studies, of single sites of lesion in the spinal cord suggestive of focal inflammatory demyelination. Recently variants of acute disseminated encephalomyelitis have been described.12 In the recurrent disseminated encephalomyelitis form of acute disseminated encephalomyelitis the first acute bout is followed by one or more episodes that reproduce all or some of the symptoms of the original attack. We are reporting three patients who satisfy the definition of recurrent disseminated encephalomyelitis.

Patients and methods

Over an eight year period three patients satisfying the criteria of transverse myelitis13 presented with acute onset complete transverse myelopathy which relapsed at the same site two to five times. The table summarises the clinical details.

All patients underwent a complete haemogram, erythrocyte sedimentation rate estimation, and collagen vascular work up (antineuclear antibody, lupus erythematosus cells, Rh factor, and anticardiolipin antibody). Blood venereal disease research laboratory, Treponema pallidum haemagglutination, fluorescent treponema antibody, HIV, and hepatitis B surface antigen tests, and a Mantoux test were carried out. Special tests in the CSF included a VDRL test and antibody studies for...
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detection of herpes simplex, herpes zoster, and cytomegalovirus infections. Two patients (1 and 2) underwent MRI of the brain and spinal cord, carried out on a 0.5 T scanner with T1, T2, and proton density weighted spin echo sequences. In patient 1, MRI was done six years after the onset of illness, during the fifth relapse. In patient 2, MRI was done five years after the onset, during the third relapse. Patient 3 underwent high volume delayed MRI during the second relapse, three years after the onset of illness. The table summarises other relevant investigations.

All patients received short courses of steroids. Recovery was defined as poor when the patient was bedridden, partial when there was dependence for activities of daily living, and good when there was complete recovery.

Results

Spinal cord MRI showed evidence of cord swelling with multiple areas of hyperintense signals on T2 and proton density scans and hypointense signals on T1 weighted images, which suggested on inflammatory demyelinating lesion. Brain MRI was normal in both the patients in whom it was done.

All patients responded to steroids. Patients 1 and 2 recovered completely after the first two attacks, subsequently the response to steroids was partial. Patient 1 recovered poorly after the last relapse, having shown no response to steroids. She had complete paraplegia with flexor spasms and had an indwelling catheter at her last follow up. Patient 2 recovered partially after the third relapse. Response to steroid was incomplete and at his last follow up he required assistance in walking and had not regained bladder control. Patient 3 has remained asymptomatic for the past three years, after completely recovering from two relapses.

Discussion

All causes of transverse myelitis have been excluded with reasonable certainty with the possible exception of viral infections, which are known to persist at specific sites in the neuraxis and cause relapsing neurological deficits. Recurring complete transverse myelopathy, in the absence of a specific aetiological diagnosis, has been variously called relapsing or recurring myelitis. These patients share several features of postinfectious associated acute transverse myelitis. Some of them had preceding fever or vaccination and all presented with acute onset ascending cord dysfunction in spinal shock. Our patients were similar except for the fact that relapse of neurological deficits always occurred at the same clinical site. In addition, CSF showed oligoclonal bands in one patient.

In one report on recurrent isolated myelitis, MRI of the spinal cord showed oedema of the cord and hyperintense signals on T2 weighted images, identical to the findings in our patients. A similar picture has also been described in MRI studies of monophasic transverse myelitis. The absence of abnormalities on brain MRI despite several relapses was an unusual finding. Even in monophasic acute transverse myelitis, which for all purposes may be considered a clinically restricted form of acute disseminated encephalomyelitis, MRI has shown silent single or multiple cerebral lesions. Thus labelling these disorders as variants of acute transverse myelitis may be incorrect, when it has been consistently shown clinically, electrophysiologically, and by MRI that there is only a single site of lesion. Despite recurrent attacks the diagnosis of clinically definite multiple sclerosis could not be applied to our patients, as there was no evidence of spatial dissemination five to eight years after the onset of disease. Isolated recurrent cord syndromes that progress to multiple sclerosis seldom resemble those of our patients. Early spatial dissemination is evident either on MRI, visual evoked potentials, or both.

The concept that acute disseminated encephalomyelitis, in adults, may progress and even recur has gained acceptance with the advent of MRI. On serial MRI, lesions have been shown to progress and new lesions have even been seen after a short interval, which may be accompanied by recurring neurological deficits. An association between the onset of relapse and the premenstrual period has been noted. A similar association with pregnancy and delivery has been reported. One of our patients had the onset of myelitis in relation to delivery.

Primary demyelinating CNS diseases constitute a broad range of disorders with monophasic acute disseminated encephalomyelitis at one end and chronic multiple sclerosis at the other. Patients such as ours would then have transitional forms of the disease. Perhaps the pattern of clinical presentation such as acute or chronic, monophasic, or recurrent forms is decided by an underlying genetic predisposition. The findings of Hillert et al. that the primary progressive form of multiple sclerosis is associated with a different HLA class 2 genotype than relapsing and remitting disease supports such a hypothesis. However, their findings have not been reproduced by others and more detailed studies are needed before this issue can be resolved.

8 Jeffrey DR, Mandell RN, Davis LE. Transverse myelitis - retrospective analysis of 33 cases, with differentiation of cases associated with multiple sclerosis and para-
Migraine

Pamela Hansford Johnson (Lady Snow), 1912-81, The humbler creation (1959)

It was true that, for her, the end of an attack was marked by involuntary weeping. These were tears she was quite powerless, with all her iron will, to check; tears scarcely of pain or of exhaustion, but of disappointment that this thing, which had tormented her for years, was never going to leave her alone, or to shorten its course by half an hour.

"Then you had it yesterday," said Maurice, sitting down on the edge of the bed. "Under control yesterday, even through that awful drive back. But I couldn't handle it this morning."

Kate had suffered from migraine headaches as a young woman, had virtually lost them during the whole of her happy marriage, had found them again a year after her husband's death. Unlike her sister, she had never been a Christian; once she had tried to be, as a resort against pain, but had found it no good. She felt some resentment towards the God in whom she did not believe because He could not, or would not, check these agonies.

She had spoken to Maurice about them often, as if clinical discussion helped. The day before an attack occurred, she often felt unnaturally well: she had come to almost dread the sense of well-being. The moment she began to see, in the air, tiny dot-and-tail phantoms like germs or tadpoles, constantly dropping down out of her range of vision and soaring up into it again, she knew that nothing could help her, that she must go through with it; but she could never keep from hoping that, just this once, she would escape.

She had confessed to Maurice, in a final weakening moment, that, for her, migraine was sometimes associated with violent sexual excitement: that was the worst thing of all. Once or twice she had attempted to ease it, only to find the attack prolonged and herself scalded by self-disgust. Indeed, there was something totally disgusting underlying this misery, something obscene in the remorseless clenching of the blood vessels, the hot tumescent in the vein, the triumphant conquest of will by agony.

Pamela Hansford Johnson (Lady Snow), 1912-81, The last resort (1956)

Mrs Baird was waiting for me after breakfast. “Would you go and see Celia? She’s got a dreadful migraine and nothing seems to touch it.” I went upstairs. The door was left open for me.

“I know” Celia said, “that this agony is my excuse for getting out of visiting Lois. She’s got a dreadful migraine and nothing seems to touch it.” I went upstairs. The door was left open for me.

It’s the left side of her face swollen and satiny, and the left eye half closed by puffiness, and watering down her face. She made me touch her temples, the glands of her neck, to see if I could feel the throbbing. “I haven’t had one of these for five years. I woke at four with this. I’ve taken every sort of poison. Hemicrania, the Greeks had a word for it.” She winced. “Pull the blind down again, will you?”

I asked her if her father could not help. “I won’t have him here, he knows that. It would kill me if he shouted, and he always does shout when I’m ill. Oh no, this is a sort of involuntary malingering. I shall feel better when you’ve got to the hospital.” She added, in a sick voice. “I told you I should improve."

I rejoined the Bairds. I was distressed and said so. “Nothing to do but forget it,” the doctor said briskly. “Its a commonplace misery and goes. It’s the penalty many people have to pay."

Rudyard Kipling, 1865-1936, In a letter to his cousin, Margaret Burne-Jones, written from Lahore on 17 June 1886

“Do you know what hemicrania means? A half headache. I’ve been having it for a few days and it is a lovely thing. One half of my head in a mathematical line from the top of my skull to the cleft of my jaw throbbs and hammers and sizzles and bangs and swears while the other half—calm and collected—takes note of the agonies next door. My discarding doctor says it’s overwork again and I’m equally certain that it rose from my suddenly and violently discarding tobacco for three days. Anyhow it hurts awfully—feels like petrifaction in sections—and makes one write abject driel."

Attributed to Alfred, Lord Tennyson, 1809-92

He gently prevails on his patients to try, The magic effects of the ergot rye.

Alexander Pope, 1688-1744, "The case of spleen (in moral essays, 1731–5)

There screen’d in shades from day’s detested glare
Spleen sighs for ever on her pensive bed, Pain at her side, and meagum at her head.


Inside my brain a dull tom-tom begins
Absurdly hammering a prelude of its own,
Capricious monotone
That is at least one definite "false note".

W S Gilbert, 1836-1911, Iolanthe (1882)

When you’re lying awake with a dismal headache and repose is taboo’d by anxiety,
I conceive you may use any language you choose to indulge in without impropriety. . . . to be continued