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Can electromyography predict the prognosis of transverse myelitis?

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Abstract The role of clinical and magnetic resonance imaging (MRI) features on the prognosis of acute transverse myelitis has been studied, but the role of electromyography (EMG) changes, although reported, has not been investigated. Seventeen patients with acute transverse myelitis were subjected to clinical evaluation, MRI scanning and concentric needle EMG. The outcome was defined on the basis of a 3-month Barthel Index (BI) score as good or poor. The EMG changes in these groups were compared. All of the patients had complete paraplegia (power grade 0), except 1 who had grade III power. Mild upper limb weakness was present in 6 patients. Joint position and vibration sense were impaired in the lower limbs, and a horizontal limit to sensory loss to pinprick was present in all of the patients. Spinal MRI was abnormal in 12 of 14 patients. EMG of the lower limb muscles in the acute stage (within 15–30 days of onset) revealed fibrillations or sharp waves or both in 11 patients. At 3-month follow-up, the lower limb power had improved in 8 and upper limbs in all 6 patients. The EMG changes also improved in 6 patients; fibrillations either disappeared or were markedly reduced. The motor unit potentials (MUPs) were of long duration, polyphasic with reduced recruitment. In 5 patients, however, no MUPs could be recorded and fibrillations persisted. Lower limb hypotonia and fibrillations on EMG were significantly related to the 3-month outcome. EMG evidence of denervation in the lower limb muscles in acute transverse myelitis suggests a poor outcome as assessed by 3-month Barthel index score.

Key words Transverse myelitis · Prognosis · Electromyography

Introduction

Acute transverse myelitis (ATM) is an acute or subacute spinal cord dysfunction characterised by paraplegia, a horizontal level of sensory impairment and sphincter dysfunction in which secondary causes such as compressive lesions, tuberculosis, syphilis, arteriovenous malformation, trauma and malignant infiltration have been excluded [4]. ATM has a uniform clinical picture, but its course is variable and prognosis difficult to predict. There have been few studies of the prognosis of ATM. In one study good, fair and poor outcomes were reported in one-third of the patients each [3]. In another study of 52 patients with transverse myelitis, those with an acute onset of symptoms had a poor prognosis [11]. Pain in the mid-thoracic region has also been regarded as a poor prognostic predictor [1]. ATM may follow an infection or vaccination, but a number of patients remain in the idiopathic group. On follow-up, 3–13.5% of patients presenting as ATM develop multiple sclerosis (MS) [7, 11]. The prognosis of ATM associated with MS is better than that of the parainfectious group. The patients with idiopathic transverse myelitis, however, had an unpredictable course and

outcome. The endpoint in the study by Jeffery et al. [5] was defined at a variable time, ranging between 1 month and 30 years. EMG changes in ATM have been reported, but their clinical significance has not been highlighted [3]. In a recent report on ten patients with ATM, the extent of signal changes on spinal cord magnetic resonance imaging (MRI), unrecordable motor evoked potentials (MEPs) (especially on lumbar stimulation) and EMG changes were associated with poor outcome [9]. In the present study, we report the EMG changes in patients with ATM and their prognostic significance.

Patients and methods

Consecutive patients with ATM based on the following criteria were included [5]: (1) acutely or subacutely developing motor sensory and sphincter disturbance, (2) spinal segmental level of sensory disturbance with a well-defined upper limit, (3) no clinical or laboratory evidence of spinal cord compression, (4) absence of other known neurological diseases such as syphilis, previously diagnosed MS, neoplasm, spinal cord arteriovenous malformation, sarcoidosis or HIV infection, (5) lack of clinical progression beyond 4 weeks.

A detailed medical history was taken, and physical examination was carried out. Muscle weakness was assessed by the Medical Research Council (MRC) scale and muscle tone by the Ashworth scale. Tendon reflexes, plantar responses, pinprick, joint position and vibration sensation were also tested. Cerebrospinal fluid (CSF) was examined for cells, protein and sugar. Full blood counts, urinalysis, erythrocyte sedimentation rate (ESR), blood sugar, serum creatinine, serum protein, serum electrolytes, collagen profile (antinuclear antibodies, anti-double-stranded (ds) DNA and rheumatoid factors), serum test for syphilis and HIV were carried out in all patients, and those with abnormal findings were excluded.

Spinal MRI or myelography was carried out in all patients. For spinal MRI, a 2.0-T superconducting system operating at 1.5 T using a flat oval surface coil was used. In three patients, MRI was carried out on a 0.5-T scanner. The entire spinal cord was imaged and T1-[500/15/3 (TR in ms/TE in ms/excitations)], proton density-(2000–2400/15–20/1) and T2-weighted (2200–2500/80–90/1) SE images were obtained in sagittal plain with slice thickness of 0.3 mm and 256 × 256 matrix.

Concentric needle EMG was carried out in all patients after 2–3 weeks from onset. Tibialis anterior, gastrocnemius, vastus medialis, biceps femoris, paraspinal, first dorsal interosseous, brachioradialis, biceps brachii and deltoid muscles were examined and selected on the basis of the clinical motor loss. Spontaneous activity, MUPs (duration, shape and amplitude) and interference pattern were evaluated. Fibrillations and sharp waves were classified from 0 to 4+ : 0 = none, + = persistent single train in at least two areas, 2+ = moderate number of fibrillations in 3 or more areas, 3+ = many in all areas, 4+ = filling the baseline in all areas [2]. For spontaneous activity a gain of 50 μV/division and for MUPs a gain of 200 μV/division with a sweep speed of 10 ms/division and filter setting 2Hz–10KHz were used. Peroneal and sural nerve conductions were also measured using the standard techniques [6].

The clinical examination and EMG studies were repeated 3 months after the onset of illness. The outcome was defined as poor or good recovery on the basis of the 3-month Barthel index score. A score of 12 or more was defined as good and below 12 as poor outcome [9]. The relationship between various clinical and EMG variables with outcome was evaluated by chi-square test.

Results

During 1993 and 1996, there were 26 patients with transverse myelopathy; these inlcuded 2 with ischaemic myelopathy, 3 with MS and 1 with herpes simplex virus I (HSV-I) myelitis in a patient with AIDS. The remaining 19 patients had idiopathic ATM. Two of these patients had a fulminant course and died from respiratory paralysis in the 1st week of illness. Detailed MRI and EMG examination in these patients was not possible. The present study is based on 17 patients with ATM. The mean age of these patients was 31.5 years (range 14–70), and 3 were female. Eight patients had a history of fever preceding their illness, and backache was present in 2. The illness peaked by 6 days (range 1–30). All patients had an acute onset of paraplegia, sensory impairment and bladder dysfunction. Muscle wasting was present in 11 patients in the lower limbs and in 1 in the small muscles of the hands as well. The wasting was noted by 3–4 weeks. All patients had complete paraplegia (MRC grade 0), except 1 who had grade III power in the lower limbs. Six patients had upper limb weakness, but it was less pronounced than that in the lower limbs. The upper limb power ranged between grade II and IV being grade II and grade III in 1 patient each, whereas in the remaining patients it was grade IV. On admission, the lower limbs were flaccid in 13, spastic in 3 and tone was normal in 1 patient. Spasticity in the upper limbs was present in 2 patients only. Knee and ankle reflexes were diminished in 12 and exaggerated in 5 patients. Upper limb reflexes were normal in 7 patients, increased in 9 and reduced in 2. Six of the patients with upper-limb reflex abnormalities also had associated weakness. The sensory loss for touch and pinprick was present in mid-thoracic level (D5-D8) in 8, upper thoracic level (D1-D4) in 4, lower thoracic region in 4 and lumbar region in 1 patient. All patients had impaired joint position sensation in the lower limbs and retention of urine.

CSF was abnormal in all patients. Mean CSF protein was 190.6 mg/dl (range 18–495), mean cell count was 59/mm^3 (range 4–310), mainly lymphocytes. Mean CSF sugar was 66.5 mg/dl (range 36–108). MRI was carried out in 14 patients and was abnormal in all except 2. The MRI abnormalities included hypo- to iso-intense signal changes in T1 and hyperintense in T2 in all except 1 who had hyperintense lesions in both T1 and T2, suggestive of haemorrhagic myelitis. The extent of signal changes ranged from normal MRI (2 patients in whom it was carried out on a 0.5-T scanner) to extensive signal changes beyond the sensory level by a mean of 8.5 spinal segments (range 2–18). The relationship between the extent of MRI signal changes (more than 10 segments) with tone ($\chi^2 = 0.36$, $df = 1$, NS) and outcome ($\chi^2 = 0.04$, $df = 1$, NS) was not found to be significant. Myelography, carried out in 3 patients in whom MRI was not possible, was normal.

Table 1 Clinical features, EMG change and outcome in patients with acute transverse myelitis (*P* paraplegia, *Q* quadriplegia)

No.	Age/sex (years)	Onset to peak (day)	Weakness	Hypotonia[b]	Sensory level	Fibrillations on initial examinations[b]	Outcome
1	14/M	2	P	+	D6	+	Poor
2	22/M	4	P	–	D8	–	Good
3	45/M	1	Q	+	D8	+	Poor
4	57/M	14	P	+	L1	+	Poor
5	15/M	1	Q	+	D4	+	Poor
6	15/F	1	P	+	D4	+	Poor
7	36/M	2	P	+	D4	+	Poor
8	17/M	15	P	–	D9	–	Good
9	25/M	1	P	–	D8	–	Good
10	52/F	2	Q	+	D2	+	Poor
11	40/M	1	Q	+	D10	–	Good
12[a]	16/M	5	Q	+	D7	+	Poor
13	33/M	1	Q	–	D7	–	Good
14	20/F	15	P	+	D10	+	Poor
15	14/M	3	P	–	D8	–	Good
16	45/M	4	P	+	D8	+	Poor
17	70/M	30	P	+	D10	+	Poor

[a] Dorsal pain
[b] + = present, – = absent

Concentric needle EMG was carried out at least 2 weeks after the onset of weakness. It revealed fibrillations and sharp waves in 11 patients in the lower limb muscles. The severity of fibrillations ranged between 2+ and 4+. The MUPs were not recordable in 5 and were of normal to short duration with reduced recruitment in the remaining patients. Upper limb EMG was normal in all except 3 patients. Upper limb denervation changes on EMG were present in 3 patients, but did not correlate with MRI signal changes in the cervical spinal cord ($\chi^2 = 0.82$, $df = 1$, NS). Sural nerve conduction velocity and amplitude of sensory nerve action potential were normal in all except 1 patient in whom sural nerve conduction was unrecordable. This patient, however, had pronounced leg oedema. Peroneal nerve conduction velocity was normal in 10 and was not recordable in the remaining patients.

All patients were followed up at 3 months after the illness. The lower limb power improved from initial grade 0 in 7 and grade II in 1 patient. The mean improvement was 3.5 MRC grades (range II–IV). Upper limb power also became normal in 5 patients, and in 1 it improved from grade II to grade III. Initially flaccid lower limbs became spastic in 2 patients; however, in 8 patients the lower limbs were persistently hypotonic. Sensation improved in 4 patients. EMG 3 months after the onset of illness revealed absence of spontaneous activity in 6 patients, and the MUPS were of long duration, polyphasic with poor recruitment. In 5 patients, fibrillations and sharp waves recorded even at 3 months, and the MUPS were unrecordable. Six patients had a good and 11 had a poor recovery. The clinical features and EMG changes in ATM patients are shown in Table 1. Hypotonia ($\chi^2 = 6.24$, $df = 1$, $P < 0.01$) and EMG changes ($\chi^2 = 12.90$, $df = 1$, $P < 0.00001$) were significantly related to the 3-month outcome, whereas the rapidity of onset, age, upper limb weakness, severity of paraplegia and backache were not. The EMG changes were present in 11 patients, and all of them had a poor outcome. All 6 patients without EMG abnormality had a good outcome. Eleven of 13 patients with hypotonia had a poor outcome whereas those without hypotonia recovered well.

Discussion

In our study, all patients with EMG changes suggestive of denervation had a poor outcome, whereas those without such changes recovered well. In ATM, the histopathological changes include varying proportions of oedema, demyelination and necrosis involving both the white and grey matter of the spinal cord. The EMG findings of denervation in the presence of normal sensory nerve conduction studies are likely to be owing to anterior horn cell involvement. In ATM, anterior horn cell involvement may suggest more severe and widespread involvement of the spinal cord with a poor chance of recovery. In myelitis, the polar forms include anterior horn cell involvement in poliomyelitis, dorsal root ganglia in herpes zoster and white matter in MS. In ATM, both grey and white matter may be involved to a varying degree. The ATM patients with predominantly white matter changes are likely to improve better and earlier compared with those with anterior horn cell involvement. The recovery following demyelination depends upon the improvement in conduction block and subsequent remyelination. The compensation following anterior horn cell damage depends upon axonal sprouting, which is so effective that up to 50% of the original number of anterior horn cells may be lost without any

clinical weakness [12]. Weakness and denervation on EMG may be owing to extensive involvement of anterior horn cells. The importance of the anterior horn cells was highlighted in an earlier study in which MS patients presenting as myelitis improved better than those with parainfectious and idiopathic ATM. Anterior horn cells may be involved in the latter, but not in MS patients. The presence of backache in the thoracic region has been reported to predict a poor outcome in ATM [1, 11]. In our study, only two patients had backache and both had a poor outcome; however, nine of our patients without any backache also had a poor outcome. Repidity of onset weakness has also been reported to be a preditor of poor prognosis, but this was not so in our study. MS patients presenting as ATM are likely to have a slower onset of illness and a better short-term outcome. A history of preceding fever before the onset of ATM (parainfectious group) has also been regarded as a poor predictor [5]. In our study, the outcome of the patients with and without preceding fever did not significantly differ. The two important prognostic predictors in our study, hypotonia and features of denervation on EMG, may be interrelated. Initial hypotonia may be owing to anterior horn cell involvement and spinal shock. In a number of our patients, it was replaced by spasticity as the stage of spinal shock passed off. A critical mass of anterior horn cells is necessary for the spasticity to manifest. Weakness, wasting and hypotonia manifest when the anterior horn cell population decreases to 10% or less. Five of our patients had persistent flaccidity, areflexia, and the EMG changes of denervation persisted even after 3 months, suggesting pronounced anterior horn cell involvement. In the remaining patients, repeat EMG revealed reduction of denervation and appearence of long-duration polyphasic potentials, suggesting reinnervation. The recruitment pattern, however, was markedly reduced, which may not only be owing to anterior horn cell loss, but also to associated spasticity.

The EMG change in ATM may be analogous to poliomyelitis in which the severity of weakness and outcome may correlate with EMG change [10]. Anterior horn cells are the final pathway, and their involvement is critical for motor deficits, even if the white matter changes are mild. In ATM, unrecordable MEPs, especially on spinal stimulation, were associated with poor outcome in our earlier study [9]. Profound loss of anterior horn cells may result in unrecordable MEPs on cortical or spinal stimulation. In some patients with anterior horn cell disease, however, marginal prolongation of central motor conduction times with reduced amplitude of MEP's has also been reported [8, 9]. Some patients who present as ATM may develop further neurological signs and turn out to have MS (3–13.5% of patients) [7, 10], although others regard this as an extremely rare possibility [5]. In our patients, the clinical and MRI features were not suggestive of MS. During this periord, only two patients with MS presented as ATM, and they have not been included in this study. The EMG features of anterior horn cell involvement may provide additional evidence against MS.

It can be conlcuded from our study that the EMG changes in ATM may provide valuable information about its prognosis. EMG, being an easily available, cheap and simple investigation, may have wider applications in the diagnosis and prognosis of the patients presenting as acute transverse myelitis.

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