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Pattern of segmental motor involvement in syringomyelia: a single fibre EMG study

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SUMMARY Single fibre EMG has been used to study the pattern of upper limb motor involvement in syringomyelia. Biceps brachii, extensor digitorum communis (EDC) and first dorsal interosseous muscles (1stDI) representing C5/6, C7/8 and C8/T1 segments, were studied. In the biceps the fibre density was slightly increased in most patients, in EDC it was about twice the normal and in the 1stDI it was about three times normal. The potentials were least stable and of longest duration in the 1stDI. These findings seem to indicate a relatively constant pattern of involvement of anterior horn cells in the brachial segments in syringomyelia.

The upper level of cavitation in syringomyelia is variable,[1] but is more frequently cervical than at any other level.[2] Indeed the disease usually begins with symptoms and signs of a lesion in the spinal cord at the cervico-thoracic junction. The location of the cavity asymmetrically within the spinal grey matter is such that neither sensory abnormalities nor weakness and atrophy always provide accurate information about the extent of the cord lesion. Nevertheless, the disease often begins with wasting of the small hand muscles,[1] an observation suggesting that the pattern of motor involvement in the cervical segments might be relatively consistent from case to case.

Electromyographic studies in syringomyelia have shown a neurogenic abnormality in the arm muscles[3,4] but, although Lenman and Ritchie[5] noted that the EMG changes were usually localized to a few segments, their distribution has not been studied in detail. In this paper we describe our observations on the pattern of motor involvement in the cervical segments in syringomyelia in ten patients, using both conventional and single fibre EMG.

Patients and methods

Ten consecutive, and thus unselected, patients with established syringomyelia were studied (table 1). Six were studied in London, (cases 1 to 6) and four in Uppsala (cases 7 to 10). All showed the typical clinical and radiological features of syringomyelia and four also had signs of syringobulbia. Their ages ranged from 30 to 68 years and symptoms had been present for 4 to 45 years (mean 20 years). There had been no progression of symptoms or signs for two years or more in five patients (cases 1, 2, 3, 7 and 8), but cases 4, 5, 6, 9 and 10 had progressed slightly in this time (table 1). In all the patients, however, progression had been most rapid in the first two or three years of the illness. In most of the patients (cases 4, 5, 7, 8 and 9) the sensory disturbance indicated a lesion located in the lower cervical and upper thoracic cord. The motor deficit in the arms suggested, similarly, that the lesion was most advanced in the lower cervical segments (table 1). In case 1, in whom there were signs of bulbar involvement, the sensory disorder extended into the upper cervical segments. All the patients except case 7 had wasted small hand muscles and five (cases 1, 2, 6, 9 and 10) had claw-hand deformities. The deltoid and periscapular muscles were relatively spared.

In each patient, conventional EMG, using concentric needle electrodes, was carried out in proximal and distal muscles of the right arm. Single fibre EMG recordings were made, using a Medelec SF25 electrode, in the right biceps brachii, extensor digitorum communis and first dorsal interosseous muscles, representing C5/6, C7/8 and C8/T1 spinal segments respectively.[6] Fibre density measurements were made in each of these muscles; the fibre density is the average number of single fibre action

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Table 1 *Clinical Data*

Case	*Age yr*	*Sex*	*Duration of symptom (yr)*	*Course*	*Distribution of impaired pin prick in right arm*	*Distribution of weakness in right arm*	*Arm tendon reflexes*	*Clinical bulbar involvement*
1	64	F	4	stable for 2 years	C2-T2 dense loss T2-L5 hypoalgesia	clawed atrophic hands severe weakness of wrist extensors biceps slightly weak deltoid normal	absent	present
2	60	F	40	stable for 10 years	C6-L2	clawed atrophic hands moderate weakness of biceps, triceps and wrist extensors	absent	present
3	65	F	45	stable for 3 years	C5-T7	weak, wasted hands biceps weak	absent	present
4	33	F	6	slowly progressive	C8-T1	weak, wasted hands weakness of flexors and extensors of wrist. Biceps and triceps strong	absent	present (cerebellar ectopia) (on myelogram)
5	30	M	19	slowly progressive	C5-C8	weak deltoids, biceps and finger extensors. Wasted hand muscles	absent	absent (cerebellar ectopia) (on myelogram)
6	64	M	30	recent progression after long remission	C6-T1	weak biceps and forearm muscles clawed, atrophic hand	absent	present
7	68	M	20	stable for 10 years	C6-C8	none	diminished	absent
8	55	M	6	stable for 2 years	C6-T1	slight weakness and atrophy of the hand	diminished	absent
9	37	M	17	slowly progressive	C2-T2	atrophic clawed hand	absent	diplopia
10	31	M	17	slowly progressive	C2-L5	atrophic clawed hand. Klippel-Feil anomaly	diminished triceps absent	absent

potentials (potential amplitude >200 μV; duration <300 μs) recorded within the uptake area of the single fibre EMG electrode in 20 different motor units in a muscle activated during weak voluntary effort.[7] Late potentials of lesser amplitude (150μV) were accepted. In wasted muscles, however, it was not always possible to record as many as 20 potentials. The upper limits of the fibre density in these muscles in normal subjects are biceps 1·7, extensor digitorum communis 1·8 and first dorsal interosseous 1·7.[8] In addition, in each of these muscles the neuromuscular jitter was determined[9] and the presence of impulse blocking was noted. Sensory and motor nerve conduction velocity measurements were made in the median and ulnar nerves in all the patients.

Results

Conventional EMG showed abnormalities in all the muscles examined. These abnormalities were most prominent in the first dorsal interosseous muscles and least in the biceps brachii. In cases 2, 4, 9 and 10 no voluntary motor unit action potentials could be recorded in the first dorsal interosseous muscles, and in case 9 no potentials could be recorded in biceps brachii. Sparse fibrillation potentials were found in the extensor digitorum communis and first dorsal interosseous muscles in all the patients, but fibrillation potentials were recorded in the biceps muscles only in cases 3 and 5. On volition, there were polyphasic motor unit action potentials of normal to moderately high amplitude, often of long duration,

Table 2 *EMG Data*

Case	Biceps				Extensor digitorum communis				First dorsal interosseous			
	FD	Duration (ms)	Number of potentials with increased jitter	impulse blocking	FD	Duration (ms)	Number of potentials with increased jitter	impulse blocking	FD	Duration (ms)	Number of potentials with increased jitter	impulse blocking
			%				%	%			%	%
1	2.1	—	0	0	2.9	—	0	0	4.3	—	2/12 (17)	2/12 (17)
2	1.6	—	0	0	3.3	—	2/18 (11)	2/18 (11)	no potentials recorded			—
3	2.4	3.5	0	0	3.2	3.8	1/20 (5)	0	5.3	6.6	2/12 (17)	0
4	2.1	2.1	1/20 (5)	0	3.9	3.3	1/20 (5)	1/20	no potentials recorded			—
5	2.3	2.6	0	0	4.1	6.4	2/20 (10)	2/20 (10)	5.8	9.2	1/12 (8)	0
6	2.7	2.9	1/20 (5)	0	4.8	5.7	3/20 (15)	2/20 (10)	5.8	4.8	5/14 (35)	3/14 (27)
7	1.2		0	0	2.4		9/19 (48)	0	2.4		7/17 (41)	0
8	1.4		0	0	2.4		6/10 (30)	1/20 (5)	2.5		5/20 (25)	2/20 (10)
9	no potentials recorded			—	4.7		7/18 (25)	4/28 (12)	no potentials recorded			—
10	1.4		0	0	4.1		2/10 (10)	2/20 (10)	no potentials recorded			—
Mean FD 1.9 (SD 0.50)					3.6 (SD 0.83)				4.4 (SD 1.43)			

FD= Fibre Density

The mean fibre densities in biceps and extensor digitorum communis are significantly different ($p<0.002$)
Similar comparison of biceps and first dorsal interosseous muscles shows a less significant difference ($p = 0.02$)

in all the patients, particularly in the extensor digitorum communis and first dorsal interosseous muscles.

Single fibre EMG recordings revealed a similar distribution of abnormality in each patient. Abnormalities were most marked in the first dorsal interosseous and least in the biceps brachii muscles (table 2). In the biceps brachii and extensor digitorum communis muscles 16 to 28 motor unit action potentials were recorded in each patient. The mean fibre density in the biceps was slightly increased (1.9). The highest fibre density found in this muscle was 2.7 (case 6). In four patients the fibre density was normal in this muscle, and in one no potentials were recorded. In the extensor digitorum communis the mean fibre density (3.6) was nearly twice that found in the biceps. In the first dorsal interosseous the mean fibre density was 4.1 (table 2). The fibre density was increased in all those first dorsal interosseous muscles in which motor unit action potentials could be recorded. In the first dorsal interossei in cases 2, 4, 9 and 10 and in the biceps muscle in case 9, no motor unit action potentials could be recorded; in these muscles, therefore, the fibre density could not be determined. In three other patients (cases 1, 3 and 5) only 12 potentials could be recorded in the first dorsal interosseous muscle, but in these muscles the fibre densities were greatly increased (table 2).

Most of the motor unit action potentials recorded with single fibre EMG in the three muscles were stable. Increased neuromuscular jitter was noted in 1% of potentials in the biceps brachii muscles examined, in 16% of potentials in the extensor digitorum communis muscles, and in 21% of potentials in the first dorsal interosseous muscles (table 2). Impulse blocking was not observed in the biceps brachii in any of the patients; it was present in 7% of motor unit action potentials in the extensor digitorum communis and first dorsal interosseous muscles (table 2). Late potentials, occurring more than 8 ms after the triggering potential, were recorded in several patients (Fig 1). These showed increased jitter and blocking more

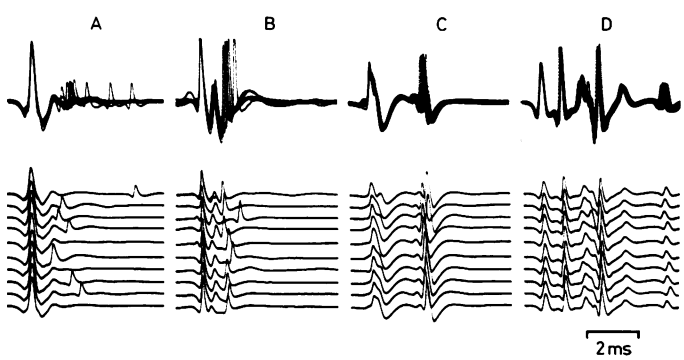

Fig 1 *Varying degrees of instability and complexity in four different recordings. A, B and C; first dorsal interosseous muscle. D; extensor digitorum communis muscle. In A a pair of single fibre action potentials is seen; the second increased jitter and blocking. In B a more complex potential, consists of 3 action potentials; the last component shows a slightly increased jitter. In D, a very complex long duration potential, with components showing a normal or slightly increased jitter, without impulse blocking. A may represent early reinnervation, and D long-standing, effective reinnervation.*

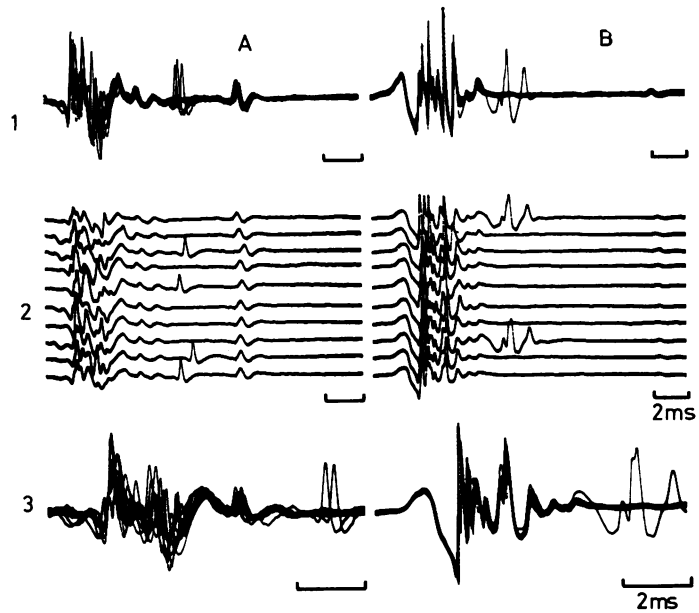

Fig 2 *Two recordings from the same first dorsal interosseous muscle. Both potentials are complex. A is an unstable potential with prominent jitter and impulse blocking. B is stable; a double discharge is seen in two of the ten sweeps.*

often than earlier components of the motor unit action potentials. The mean duration of the recorded motor unit action potentials was most increased in the first dorsal interosseous muscles and least abnormal in the biceps brachii muscles. In the first dorsal interosseous and extensor digitorum communis muscles most patients were unable to maintain a steady innervation rate. In a number of the recordings double discharges were seen (fig 2). These discharges appeared 5 to 10 ms after the initial component. Their shape was usually different from the triggering motor unit action potential complex.

Sensory and motor nerve conduction velocities in the median and ulnar nerves were normal in all patients in whom measurement was possible. In cases 9 and 10 motor nerve conduction velocity could not be measured in the ulnar nerve, because no muscle action potential could be recorded in the abductor digiti minimi.

Discussion

In neurogenic disorders an increased fibre density indicates that reinnervation has occurred. Because the technique requires only slight activation of a muscle, and type 1 units are generally first activated,[10] the motor unit action potentials recorded with single fibre EMG are predominantly type 1 motor units.[11] Indeed, an increased fibre density has been shown to correlate with histochemical evidence of type 1 fibre grouping in muscle biopsies.[12] The abnormalities found with single fibre EMG in our patients with syringomyelia, consisting of increased fibre density, and of some potentials with increased neuromuscular jitter and impulse blocking, are similar to those found in other disorders affecting anterior horn cells, particularly the slowly progressive form of motor neuron disease.[13] The combination of complex but stable motor unit action potentials, with an increased fibre density, is evidence of a chronic disorder with effective reinnervation. Our single fibre EMG measurements provide quantifiable evidence of abnormality, particularly of increased fibre density, even in muscles such as the biceps brachii, in which conventional EMG was virtually normal. In slowly progressive disorders, however, a stage is eventually reached at which few motor units remain and severe neurogenic atrophy has occurred. At this stage it may be difficult or impossible to record functioning motor units, as in the first dorsal interosseous muscles in our cases 2, 4, 8 and 9.

In our patients the neuromuscular jitter was increased to a similar degree both in those in whom the disease had progressed in the previous two years, and in those in whom it had been stable during this period (table 3). However, in the group of patients with recent progression impulse blocking was more frequent (13% in the extensor digitorum communis) than in those with a stable course (3% in the extensor digitorum communis). Impulse blocking is due to failure of excitation of a muscle fibre either at the neuromuscular junction or in the terminal

Table 3

Clinical	Case	Exterior digitorum communis			First dorsal interosseous			Duration of disease (yrs)
		FD	Neuromuscular jitter (%)	Impulse blocking (%)	FD	Neuromuscular jitter (%)	Impulse Blocking (%)	
Stable	1	2.9	0	0	4.3	17	14	4
course	2	3.3	11	11	no potentials recorded			40
	3	3.2	4	0	5.3	17	0	45
	7	2.4	48	0	2.4	41	0	20
	8	2.4	30	5	2.5	25	10	6
		—	—	—	—	—	—	—
	mean	2.8	17%	3%	3.5	25%	6%	23 yrs
Progressive	4	3.9	5	5	no potentials recorded			6
course in	5	4.1	10	10	5.8	5	0	19
last 2	6	4.8	15	15	5.8	35	22	30
years	9	4.7	25	25	no potentials recorded			17
	10	4.2	10	10	no potentials recorded			17
		—	—	—	—	—	—	—
	mean	4.2	13%	13%	5.8	20%	11%	18 yrs

In both extensor digitorum brevis and first dorsal interosseous muscles the fibre densities in patients with stable and progressive course were significantly different ($p < 0.002$).

nerve branches.[14] This functional abnormality probably occurs both in degenerating axons and in immature regenerating axons and neuromuscular junctions. Impulse blocking is therefore associated with weakness and fatiguability. The neuromuscular jitter was increased both in the clinically progressive, and in the stable groups of patients. This phenomenon, when it occurs without blocking, is *not* associated with weakness or fatiguability. The fibre density in the extensor digitorum communis muscle was greater ($p < 0{\cdot}002$: table 3) in the group of patients (mean 4·1) with a progressive course than in the group with a stable course (mean 2·8). Similar trends were noted in the first dorsal interosseous muscles.

In a previous study of three patients with recently progressive syringomyelia.[13] a similar increase both in fibre density and in impulse blocking was observed in the extensor digitorum communis muscle. The combination of an increased fibre density, and increased frequency of impulse blocking in the group of patients with a progressive course is consistent with a continuing loss of anterior horn cells, leading to marked peripheral axonal sprouting, as shown histologically in methylene blue studies of the terminal innervation pattern.[15] In our patients these single fibre EMG abnormalities were not related to the duration of symptoms and it may therefore be inferred that a moderate increase in fibre density ($> 3{\cdot}5$) with infrequent impulse blocking, indicates that the syringomyelia is in a stable phase.

In neurogenic disorders, including syringomylia, increased motor unit action potential duration could be due to several factors, including dispersion of the motor end-plate zone, slowed propagation velocity in small or reinnervated muscle fibres, and slowed conduction in thin, sprouting axons,[13 16] Low discharge rates and irregular firing patterns of motor units have been reported with upper motor neuron lesions,[17] but in our patients it seems likely that the predominant disorder was lower motor neuron in type, from direct damage to anterior horn cells, although descending projections to these cells could have been interrupted locally by the syrinx.

In our patients the abnormalities found were most prominant in the first dorsal interosseous muscles (C8/T1 segments) and least in the biceps brachii muscles (C5/6 segments). This pattern of segmental involvement differs from that found in motor neuron disease, in which the abnormality is less constantly distributed[18] and in which the motor unit action potentials are generally less stable than those we have recorded in syringomyelia[13]. Although the pattern of segmental involvement in syringomyelia is consistent from case to case, this pattern of abnormality could also result from other disorders, such as cervical radiculopathy and cervical intramedullary neoplasm. In cervical radiculopathy other EMG investigations such as F response studies[19] are usually abnormal. Further, sensory nerve conduction studies may be abnormal in radiculopathies but are normal in syringomyelia.[20]

The constant pattern of motor involvement found in our patients, despite the varied duration and severity of their disease, suggests that the pattern of segmental damage to anterior horn cells in syringomyelia may be similar from case to case. Pathological studies of the distribution of anterior horn cell damage in syringomyelia are not available[21] and most recent reports have been concerned

with associated craniovertebral[1] or other anomalies.[2] Our EMG data suggests that detailed pathological study of the pattern of involvement of the ventral grey matter of the spinal cord, and of the distribution of cavitation in the cord, might be interesting.

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