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ORIGINAL COMMUNICATION

Distal neuralgic amyotrophy

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Abstract Neuralgic amyotrophy consists of severe pain around the shoulder and arm followed by weakness in one or several muscles of the same area. We describe four patients with distal neuralgic amyotrophy in whom acute, severe, and transient pain around the shoulder or arm was followed by weakness of the forearm and hand muscles only. Minor sensory symptoms were present in only one patient. The presence of structural lesions causing the extent of the forearm and hand motor deficit was excluded by ancillary examinations. Electrophysiological studies showed a motor axonopathy and minimal sensory axonopathy. A follow-up of 2 years or longer showed either spontaneous improvement or residual motor deficit. Unfamiliarity with a clinically distal localization of neuralgic amyotrophy may result in misdiagnosis of lower cervical (poly)radiculopathy in view of the distal localization of the motor deficit and the high prevalence of coincidental abnormalities of the lower cervical spine on plain radiography, computed tomography, or magnetic resonance imaging.

Key words Neuralgic amyotrophy · Brachial plexus neuropathy · Muscle atrophy

Introduction

Neuralgic amyotrophy (NA) and idiopathic brachial plexus neuropathy refer to a syndrome consisting of acute, severe, and transient pain in the shoulder, arm, or both, followed by flaccid weakness in the same area and less prominent or absent sensory symptoms [2, 4, 6, 13, 15, 18, 21, 23]. The term NA has the advantages of (a) stressing the discrepancy between major motor deficit and minor or even no sensory signs and (b) not limiting the site of the lesion to the brachial plexus [7]. A distal form of NA (NAd) in which weakness is limited to the forearm and hand muscles is very uncommon and not explicitly mentioned in many neurological textbooks. As this may contribute to diagnostic delay, the characteristics of NAd based on four personal cases and data from the literature are outlined.

Methods and patients

All patients who had been referred between August 1993 and July 1995 to our neurological outpatient clinic for acute brachialgia followed by weakness of one or several muscles of the forearm, the hand, or both were considered for a possible diagnosis of NAd. Severity of weakness was assessed using the Medical Research Council (MRC) five-point scale, ranging from grade 0 (paralysis) to grade 5 (normal strength). We used the Mayo Clinic's segmental distribution scheme for determining the extension of motor deficit and the Mayo nine-point scale for assessing the reflex responses, ranging from –4 (absent) to +4 (maximal exaggeration) [14].

Electrophysiological assessment consisted of needle electromyography (EMG), measurement of the motor nerve conduction velocities (MCV), sensory nerve conduction velocities (SCV), the amplitude of the compound muscle action potentials (CMAPs), and sensory nerve action potentials (SNAPs). CMAPs, distal motor latencies, and F waves along the median and ulnar nerves were recorded following stimulation at the wrist. SNAPs were recorded by ring electrodes wrapped around the first digit (median and radial nerve) and the fifth digit (ulnar nerve) following antidromic stimulation at the wrist. The median and ulnar MCVs were assessed in the

forearm; the sites of stimulation for calculating the median and ulnar MCVs were the elbow and the wrist. The CMAPs and F waves were recorded by surface electrodes from the abductor pollicis brevis (median nerve) and the abductor digiti quinti (ulnar nerve). Fibrillation potentials and positive sharp waves in a particular muscle were graded in terms of four categories (from +1 to +4) according to Kimura [11]. Nerve conduction studies were performed with limb temperatures of at least 32 °C. Imaging of the cervical spine and the cervicobrachial plexus consisted of plain cervical radiography, magnetic resonance imaging (MRI), and in some cases computed tomography (CT) myelography.

Clinical inclusion criteria for diagnosing NAd were (a) sudden and severe pain in the shoulder, the arm, or both, progressively decreasing within a few weeks; (b) flaccid weakness confined to several muscles of the forearm and the hand occurring within 2 weeks after the pain had started; (c) stabilization or (partial) recovery of weakness and atrophy during a follow-up period of at least 2 years.

Exclusion criteria were (a) persistence of substantial pain over a period of more than 4 weeks; (b) further progression of weakness or the occurrence of other symptoms in the involved area during the follow-up period of 2 years; (c) the presence of a systemic disorder, malignancy, or another disease that might be the cause of the neurological signs; (d) structural lesions on MRI, CT myelography, or both, to which the motor deficit could be attributed.

A *follow-up* period of at least 2 years was required to confirm either stabilization or (partial) recovery of the motor deficit.

Between August 93 and July 1995 four patients (three men) fulfilled all inclusion and exclusion criteria. Their age ranged from 44 to 70 years. The clinical characteristics and the results of EMG are described. Results of measurements of MCVs and SCVs are presented in Table 1.

Table 1 Nerve conduction studies. (*SNAP* Sensory nerve action potential, *SCV* sensory conduction velocity, *CMAP* compound muscle action potential, *DML* distal motor latency, *MCV* motor conduction velocity, *NR* not recordable, *ND* not done, >: lower limit, < upper limit of normal values)

Nerve	Normal	Patient 1	Patient 2	Patient 3	Patient 4
SNAP					
Median (μV)	> 10	20	10	50	ND
Ulnar (μV)	> 8	6[a]	10	50	ND
Radial (μV)	> 4	3.5[a]	5	16	ND
SCV					
Median (m/s)	> 41	45	46	56	ND
Ulnar (m/s)	> 40	65	48	54	ND
Radial (m/s)	> 42	56	54	57	ND
CMAP					
Median (mV)	> 5	0.2[a]	NR	3[a]	1.0[a]
Ulnar (mV)	> 7	7[a]	0.2[a]	11	0.6
DML					
Median (ms)	< 4.0	3.9	NR	3.5	3.0
Ulnar (ms)	< 3.4	3	3.9[a]	2.3	2.4
F wave latency					
Median (ms)	*	ND	NR	25	30[a]
Ulnar (ms)	*	31	ND	24	30[a]
MCV					
Median (m/s)	> 49	47[a]	NR	62	62
Ulnar (m/s)	> 49	57	34[a]	53	49

*F wave latency: normal values depend in the patient's height

Case reports

Patient 1

A 64-year-old man experienced sudden and severe pain in the left forearm radiating to the fingers 1–4. There was no pain in the neck or the upper arm. His history was uneventful. Clinical examination showed diffuse weakness of the hand flexors and the intrinsic hand muscles. There was no sensory loss. The pain disappeared within 2 weeks. MRI of the cervical spine showed slight compression of the left C6 and right C7 nerve roots due to spondylotic narrowing of the intervertebral foramina. Four months later he was referred to a neurosurgeon for a left C5-C6 foraminotomy. The neurosurgeon's diagnostic doubts led to a second opinion.

Five months after the first symptoms there was still diffuse weakness of the forearm and hand muscles with the following MRC grades: intrinsic hand muscles 3–4, finger flexors and flexor carpi ulnaris 3, thumb and finger extensors 4, and extensor carpi ulnaris 4+. There was a diffuse atrophy of the forearm and hand muscles. Sensory symptoms or signs were not found. The left triceps brachii stretch reflex was –2 and the right one was 0. Five months after the first symptoms EMG showed abundant fibrillation potentials and positive sharp waves in the following muscles of the left arm and hand: abductor digiti quinti (+2), abductor pollicis brevis (+2), flexor carpi radialis (+3), flexor carpi ulnaris (+1), pronator quadratus (+3), and triceps brachii (+ 1). Sporadic fibrillation potentials (+1) were seen after needle insertion in the left paracervical muscles at the level of C5-C6 and C6-C7. Results of nerve conduction studies are listed in Table 1. NAd was diagnosed on the basis of the clinical and EMG data showing motor deficit extending beyond the territories of one cervical nerve root or one peripheral nerve. After 2.5 years the distal weakness had only slightly improved. No other neurological symptoms occurred.

Patient 2

A 63-year-old painter presented to the emergency room of the nearest hospital because of acute excruciating pain in the left shoulder radiating to the left forearm and hand. A tentative diagnosis of acute spondylotic cervical radiculopathy was made on the basis of marked spondylotic changes on radiograms of the cervical spine. One week later the patient noted substantial weakness of the left forearm and hand muscles. The pain subsided within 2 weeks, but the weakness persisted. We examined this patient 3 months after the brachialgia had started and found diffuse weakness grade 2–3 and atrophy of the forearm and hand muscles. There was also minimal weakness (grade 4+) of the biceps and triceps brachii muscles. The strength of the shoulder musculature was normal. There were no sensory changes.

EMG 3 months after the onset of weakness showed marked fibrillation potentials and positive sharp waves in the left abductor pollicis brevis (+3), interosseus primus (+3), flexor carpi radialis (+3), and biceps brachii (+2); the triceps brachii muscle showed no evidence of denervation potentials. The CMAPs showed very low amplitudes in several hand muscles, which contrasted with the normal SNAPs. Radiograms of the cervical spine showed spondylotic changes, most pronounced at the levels C5-C6 and C6-C7. MRI of the cervical spine and the cervicobrachial plexus showed slight bilateral foraminal compression of the C7 nerve roots. Tests of serum for syphilis and *Borrelia burgdorferi* infection were negative. CSF analysis was not carried out. Clinical reexamination 2 and 3 years after the acute episode showed the same flaccid weakness and atrophy of the forearm and hand muscles (Fig. 1). There was no sensory loss.

Patient 3

A 44-year-old nurse experienced sudden and severe right-sided shoulder and upper arm pain followed by increasing weakness of

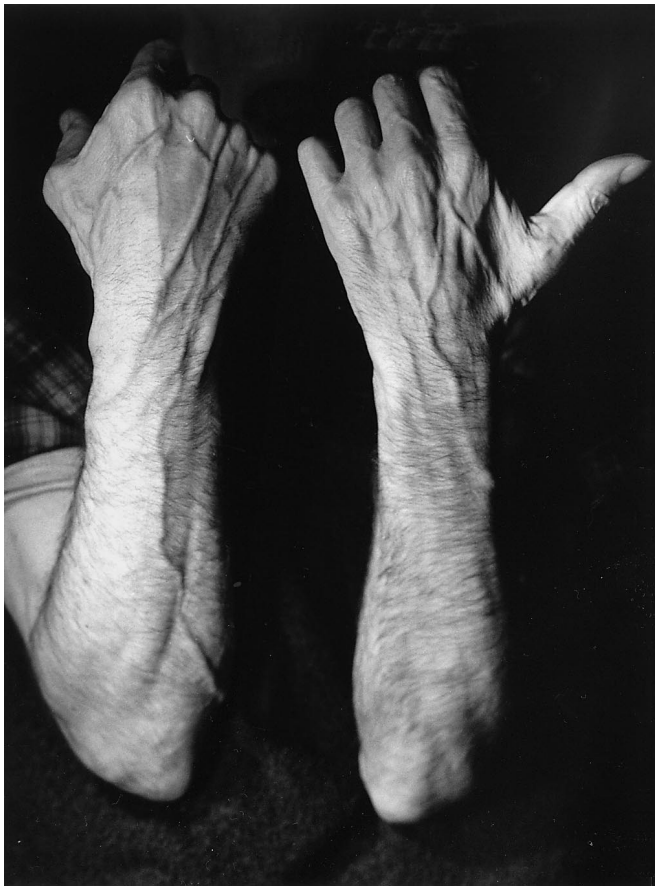

Fig. 1 The left forearm and hand of patient 2 showing severe and diffuse muscle atrophy. There was no sensory deficit

the intrinsic hand muscles with impaired flexion and extension of the fingers, most pronounced in the fingers 3–5. She had no sensory symptoms. Clinical examination showed weakness and slight atrophy of the hand flexors and extensors grade 4 and slight weakness (grade 4+) of the opponens pollicis. EMG showed fibrillation potentials and positive sharp waves in the right abductor pollicis brevis (+3), abductor digiti quinti (+2), flexor carpi radialis (+1), and extensor indicis proprius (+1). The MCVs along the median and ulnar nerves were within normal ranges, as were the SCVs along the radial, median, and ulnar nerves (see Table 1). MRI of the cervical spine and the cervicobrachial plexus showed no abnormalities.

Additional investigations showed no evidence of systemic disease or malignancy. Three months later complete recovery was observed. There was no recurrence of pain or weakness during a follow-up period of 40 months.

Patient 4

A 50-year-old accountant experienced acute pain and muscle cramps in the left shoulder and arm followed by impaired extension of the 4th and 5th fingers of the left hand after 1 week. He also complained of intermittent tingling sensations at the ulnar side of the left forearm and hand. The weakness rapidly extended to the hand and finger extensors and the intrinsic left hand muscles. CT myelography showed a slight left-sided disc protrusion at the C5-C6 level and minor degenerative changes with a questionable left C6 nerve root compression. The patient was referred to a neurosurgeon who referred the patient for additional investigations. Two months after the first symptoms we found diffuse weakness of the forearm and hand muscles with the following MRC grades: triceps brachii 4, extensor carpi radialis 3, extensor carpi ulnaris 2, finger extensors grade 1, intrinsic hand muscles varying between 0 (abductor digiti quinti) and 2 (flexor pollicis brevis), and finger flexors 4. There was diffuse atrophy of the left forearm and hand muscles. Minimal and inconsistent hypesthesia was found in dermatomes C7 and C8.

EMG showed marked fibrillation potentials and positive sharp waves in the flexor pollicis brevis (+3), the abductor digiti quinti (+3) and the extensor digitorum communis (+3). There were no abnormal findings in the paracervical, shoulder and upper arm muscles. MCVs were normal except for minimal slowing along the ulnar nerve (see Table 1). SCVs were not assessed. Ancillary investigations showed no evidence of local or systemic disease or malignancy. After 2 years the forearm and hand muscle weakness remained unchanged; no other neurological symptoms had occurred.

In all patients analyses of hematology, blood chemistry, and urine showed no relevant abnormalities; chest radiograms were normal.

Discussion

Our patients presented clinically with idiopathic NAd consisting of acute and severe pain in the shoulder and arm, followed by weakness restricted to the forearm and hand muscles and only minor or even no sensory loss. A follow-up during at least 2 years showed that the residual motor deficit varied from severe weakness to complete recovery. The electrodiagnostic investigations showed a similar pattern as commonly described in NA and brachial plexus neuropathy [1, 5, 8, 19, 21, 26, 27] consisting mainly of polysegmental fibrillation potentials, positive sharp waves, reduced CMAPs, normal or minimally slowed MCVs, normal SCVs, and either normal or minimally reduced SNAPs. These findings were consistent with a predominantly motor axonopathy either in the territory of the brachial plexus or in that of several nerves within the arm [8]. In patient 2, with severe motor deficit, the motor axonopathy was associated with slowed MCV suggesting secondary demyelination. Minor EMG abnormalities in the clinically unaffected myotome C6 contributed to the demonstration that muscle involvement was consistent with either a brachial plexopathy or a multiple brachial mononeuropathy. Although in patient 1 some paracervical fibrillation potentials were registered, we did not find MRI abnormalities explaining the extension of the motor deficit. The most plausible hypothesis is that the paraspinal fibrillation potentials were a coincidental finding related to spondylotic narrowing of the intervertebral foramina with subsequent nerve root compression.

The weakness in NA is frequently limited to one or several muscles of the shoulder or the upper arm [15, 23]. Involvement of the entire brachial plexus including the forearm and hand muscles has been reported in 10–40% of the patients [6, 7, 12, 15, 21, 25, 26], but weakness limited to the forearm and hand muscles seems very uncom-

mon [5, 7, 12, 19, 20]. Magee and De Jong's [13] series of 23 patients with paralytic brachial neuritis contained no single patient with selective involvement of the lower brachial plexus. In their series of 136 patients with NA Parsonage and Turner [15] described five persons who had a combination of proximal and distal weakness and only one patient with NAd. Among their 99 patients with brachial plexus neuritis, Tsairis and Dyck [21] found only eight in whom there was selective involvement of the lower brachial plexus with predominance of motor signs. In our patients we did not find the predilection for motor deficit in the territory of the radial or anterior interosseus nerves as observed by others [3, 9, 10, 16, 17, 19, 22, 24]. Most patients with NAd show a marked predominance of motor involvement as seen in proximal NA [5, 7, 12]. England and Sumner [7] illustrated in a selected series of nine NA patients that the clinical presentation of neuralgic amyotrophy may be very diverse and that the site of the lesion(s) in some patients is probably localized in one or more nerves of the arm rather than in the brachial plexus.

In two of our patients a misdiagnosis of cervical (poly)radiculopathy had initially been made; this was probably due to the combination of severe arm pain, distal localization of the motor deficit and the presence of coincidental degenerative abnormalities of the cervical spine. We conclude that NAd should be considered in patients with severe brachialgia and weakness confined to the forearm or hand muscles especially when cervical abnormalities shown on MRI or CT myelography are not extensive enough to explain the distribution of the neurological deficit.

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