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Clinical Article

Neuroradiologic and Neurophysiologic Findings of Neuralgic Amyotrophy

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Objective : Neuralgic amyotrophy (NA) is a distinct clinical syndrome that is characterized by the acute onset of shoulder and arm pain, weakness, and sensory loss. The purpose of this study was to assess the clinical characteristics of NA and to determine appropriate diagnostic modalities.

Methods : We reviewed the medical and radiologic records of 10 patients diagnosed with NA retrospectively. Neurophysiologic studies were performed in all patients and magnetic resonance neurography was performed in the last three patients.

Results : A total of 10 patients were enrolled in our study. All patients had clinical findings compatible with NA. The most common clinical presentation was severe shoulder pain and weakness in seven patients (70%). Neurophysiologic study results were abnormal in all patients. Brachial plexus magnetic resonance neurography showed that the affected brachial plexus showed a thickened and hyper-intense trunk. All patients were managed conservatively with analgesics and physical therapy. The pain and paralysis of all patients improved clinically within 6 months of the initiation of treatment.

Conclusion : NA is a rare disease but the symptoms of NA can mimic those of other diseases. Neurophysiologic studies and magnetic resonance neurography are extremely useful tools for the diagnosis of NA.

KEY WORDS : Neuralgic amyotrophy · Brachial plexus · Diagnosis · Neurophysiology · Magnetic resonance imaging.

INTRODUCTION

Neuralgic amyotrophy (NA), also known as idiopathic brachial plexopathy or "Parsonage-Turner syndrome", is a distinct clinical syndrome of unknown etiology that involves the brachial plexus. This syndrome is characterized by the acute onset of shoulder and arm pain followed by motor weakness and sensory loss[6,11,15,19].

It is a rare condition and its incidence is approximately 1.64/100,000/year[15]. Precipitating factors include infectious diseases, connective tissue diseases, immunizations/medications, pregnancy, and surgical operations; however, approximately 54% of patients have no known precipitating factors[10,11,15,19].

Diagnosis is based on the patient's medical history, clinical symptoms, and physical examination results[14,22]. The most helpful diagnostic modalities are neurophysiologic and neuroradiologic studies[5,7,10,14]. However, the initial symptoms of NA are very similar to the symptoms of other neurological diseases and shoulder disorders. NA may therefore be misdiagnosed as a cervical spine disease such as acute cervical disc disease, or as a shoulder disorder such as a rotator cuff tear[6,15,21]. Occasionally, patients who are misdiagnosed may receive unnecessary cervical spine or orthopedic operations[15]. To avoid unnecessary diagnostic procedures or surgical interventions, it is important to distinguish NA from other alternative diseases. The purpose of this study was to assess the clinical characteristics and neurophysiologic and neuroradiologic findings of NA.

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MATERIALS AND METHODS

We retrospectively reviewed the medical records of all patients who were examined between 2006 and 2009 in our department and who had clinical evidence of acute brachial plexopathy. We excluded patients with predisposing factors such as trauma, infection, radiation injury, or tumors. Clinical parameters such as age, sex, symptoms, and neurologic

deficits were reviewed. Pain severity was assessed by a visual analogue scale (VAS). Neurophysiologic studies were performed in all patients 3 weeks after their initial presentation to avoid any false negative result. The last three patients were evaluated by magnetic resonance neurography (MRN) using a 1.5-T scanner (Gyroscan Intera, Philips Medical Systems, Best, The Netherlands). Coronal short tau inversion recovery (STIR) imaging was obtained at TR/TE 5000.00/52.00. Data were analyzed using the software package SPSS (version 12.0 for Windows, SPSS Inc, Chicago, IL, USA). The Wilcoxon signed rank test was used for statistical analyses. *p*-values below 0.05 were considered to be significant.

RESULTS

Population

A total of 10 patients (8 males and 2 females) were diagnosed with NA based on positive neurophysiologic findings (Table 1). The mean age was 55.7 ± 16.9 years (range 18-71 years). Three patients had hypertension, one had diabetes mellitus, and one patient had immunoglobulin A nephropathy-induced chronic renal failure. The latter patient had been receiving hemodialysis three times per week for the past 7 years.

Clinical features

Clinical symptoms were present on the right side in six patients and on the left side in four patients. The most common clinical presentation was severe shoulder pain and weakness in seven patients (70%), finger weakness in two patients (20%), and anterior chest wall pain and shoulder weakness in one patient (10%). The mean initial VAS score was 6.6 ± 1.3 (range 4-8). The duration of pain was 2.9 ± 3.3 weeks (range 1-12 weeks). Only four patients (40%) were first diagnosed with NA when they visited our hospital, while the remaining six patients were misdiagnosed as follows. Three patients who had shoulder pain and weakness were misdiagnosed with cervical disc herniations. One patient who presented with shoulder pain and anterior chest wall pain was suggested to have pancoast syndrome. The other two patients, who presented with mild to moderate pain and definite weakness of the 4th and 5th fingers, were diagnosed with ulnar nerve problems including cubital tunnel syndrome.

Cervical spine magnetic resonance imaging (MRI) was performed in four patients to diagnose or rule out degenerative cervical spinal disorders. One patient had a non-specific finding, but the other three patients had abnormal findings with mild to moderate disc herniations. MRI imaging of the second patient, who had right-sided shoulder pain and weak-

Table. 1. Clinical manifestations of NA

Patient No.	Age (years)	Sex	Initial presentation	Initial impression	Side	Involved muscle and motor power	Sensory changes
1	42	M	SP and SW	CDH	Left	Biceps (Gr IV) Wrist ext (Gr III) Interossei (Gr III)	Patchy
2	69	M	SP and SW	NA	Right	Deltoid (Gr II) Biceps (Gr II)	Patchy
3	51	M	Finger weakness	CuTS	Left	4,5 finger E (Gr II) Interossei (Gr III)	ulnar side
4	71	M	SW Anterior chest wall pain	Pancoast syndrome	Right	Deltoid (Gr III) Biceps (Gr IV)	None
5	61	M	SP and SW	NA	Right	Deltoid (Gr III) Biceps (Gr III) Brachialis (Gr III)	None
6	60	M	SP and SW	CDH	Left	Deltoid (Gr II) Biceps (Gr III)	2nd and 3rd fingers
7	46	F	Shoulder and upper arm pain and weakness	CDH	Right	Deltoid (Gr II) Biceps (Gr II)	None
8	18	M	SP and SW	NA	Right	Deltoid (Gr III) Biceps (Gr II) Triceps (Gr IV-)	None
9	68	M	SP and SW	NA	Left	Deltoid (Gr II) Biceps (Gr III) Triceps Gr III)	Patchy
10	71	F	Finger weakness	CuTS	Right	4, 5 finger E (Gr II) Interossei (Gr III)	Ulnar side

No.: number, SP: shoulder pain, SW: shoulder weakness, NA: neuralgic amyotrophy, CDH: Cervical disc herniation, CuTS: cubital tunnel syndrome, Gr: grade , E: extensor

ness, revealed a protruding disc at the C4/5 level on the right side (Fig. 1). This patient was strongly suspected to have cervical disc herniation, but the deltoid and biceps weakness were too severe for definite diagnosis (grade II), and the sensory changes also had an atypical distribution. We therefore diagnosed this patient with NA based on neurophysiologic studies performed 3 weeks after this patient was first examined.

Examination of muscle function revealed that the most commonly affected muscle was the biceps (eight patients) followed by the deltoid (seven patients), palmar interosseous (three patients), triceps (two patients), finger extensor (two patients) and brachioradialis (one patient) (Table 1). The mean initial motor power was grade 2.4 ± 0.6 (range grade II-IV). Six patients had atypical sensory changes, but four patients had no sensory symptoms (Table 1).

Neurophysiologic studies

Neurophysiologic studies were performed in all patients 3 weeks after initial presentation. Nerve conduction study (NCS) findings were reduced compound muscle action potential (CMAP) in five patients, prolonged CMAP in three patients, and reduced sensory nerve action potential (SNAP) in three patients. Nerves involved in NCS were the axillary nerve in seven patients, radial nerve in two patients, medial nerve in two patients, and the ulnar, medial cutaneous, lateral cutaneous, and antebrachial nerve in one patient each (Table 2). The most commonly involved trunk of the brachial plexus was the upper trunk in seven patients, while the remaining three patients had lower trunk involvement.

Magnetic resonance neurography findings

The last three patients were diagnosed with brachial plexus MRN. MRN was taken at the acute phase (within 3 weeks in all three patients). MRN imaging of these patients showed a thickened and hyperintense trunk of the affected brachial plexus, consistent with plexitis (Fig. 2).

Treatment and clinical outcomes

The mean follow-up period was 7.8 ± 3.8 months (range 6-18 months). All of the patients were managed conservatively with analgesics and physical therapy. At the last examination, the average VAS score was 2.2 ± 1.0 (range 1-4) and the severity of pain was significantly improved compared with the initial examination (p value < 0.05) (Table 3).

The mean motor power had improved to grade 4.5 ± 0.7 (range III-V) (p value < 0.05). In half of the patients, motor weakness was improved within 2 months, but in some patients, motor weakness lasted for up to a year. Six patients experienced no weakness after 6 months, while four patients still experienced weakness at 6 months (fair to good).

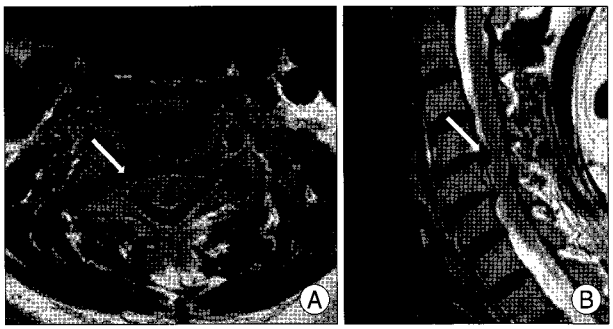

Fig. 1. Sagittal (A) and axial (B) T2-weighted MRI images of patient No. 2 show a right-sided protruded disc (white arrow) at the C4/5 level and compression of the right C5 nerve root by the protruded disc.

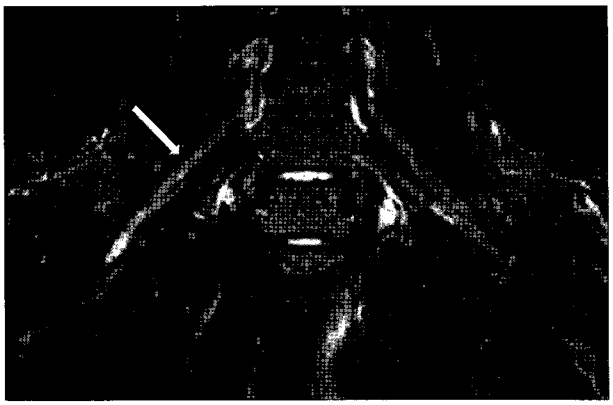

Fig. 2. STIR images of the MRN of patient No. 8 shows a diffuse thickening and increased signal intensity in the region of the right upper trunk of the brachial plexus (white arrow).

Table 2. Neurophysiological findings of NA

Patient No.	NCS findings	Results
1	Prolonged CMAP : radial nerve	LTP
	Reduced SNAP : lateral cutaneous branch	
2	Prolonged CMAP : median and axillary nerve	UTP
3	Reduced CMAP : radial and ulnar nerve	LTP
4	Prolonged CMAP : axillary nerve	UTP
5	Reduced CMAP : axillary nerve	UTP
6	Reduced SNAP : axillary nerve	UTP
7	Reduced CMAP : axillary nerve and MC nerve	UTP
8	Reduced CMAP : axillary nerve	UTP
9	Reduced CMAP : axillary nerve	UTP
10	Reduced SNAP : antebrachial nerve	LTP

No. : number, NCS : nerve conduction study, CMAP : compound muscle action potential, SNAP : sensory nerve action potential, MC : medial cutaneous, UTP : upper trunk plexopathy, LTP : lower trunk plexopathy

DISCUSSION

NA was first described by Parsonage and Turner in 1948, and has been variously referred to as "multiple neuritis of the shoulder girdle", "localized neuritis of the shoulder girdle", "infective neuritis", and "acute brachial radiculitis"[11]. Parsonage and Turner[11] reported that there was evidence of some precipitating factor in 98 of the 136 cases they examined. Preci-

Table 3. Clinical outcomes

Patient No.	Initial SOP (VAS)	Last SOP (VAS)	DOP (weeks)	Initial LMP (Grade)	Last LMP (Grade)
1	7	3	3	IV	V
2	8	4	3	II	V
3	4	1	12	II	IV
4	6	1	2	III	V
5	7	2	1	III	IV
6	8	3	2	II	V
7	5	2	1	II	V
8	7	1	1	II	V
9	8	3	1	II	III
10	6	2	3	II	IV

SOP : severity of pain, DOP : duration of pain, LMP : lowest motor power

pitating factors include trauma, infection, previous operations, medications, neoplasms, and thoracic outlet syndrome[10,11,15,19]. Trauma is the most common cause of brachial plexopathy, while the most common cause of non-traumatic brachial plexopathy is idiopathic brachial neuritis, namely NA[10].

To diagnose NA, medical history taking and physical examination are critical[14]. The classic triad of NA symptoms is pain, weakness, and sensory loss[6,11,15,19]. The syndrome starts with the acute onset of a severe and relentless pain in the shoulder. Pain is not limited to the shoulder and may spread to the scapula, arm, forearm, elbow, or finger region[15,21]. The causes of pain are damaged nerves in the plexus and scapular instability caused by a paresis of the serratus anterior and rhomboids or trapezius[21]. Pain may persist for several hours to weeks[11,15]. In our study, all patients had moderate to severe pain in the acute phase that involved the shoulder, forearm, fingers, and/or chest wall. Pain persisted for an average of 2.9 ± 3.3 weeks (range 1-12 weeks), consistent with previous reports.

The motor and sensory symptoms of NA are patchy; this is an important indication of NA[21]. Paresis may occur within the first 2 weeks after the initial presentation[15]. Paresis involves the brachial plexus, including the long thoracic nerve and anterior interosseus nerve, and can sometimes affect the phrenic nerve or lower cranial nerve[2,9,13,22]. In most patients, improvement in motor power begins between the first and sixth months[22]. In our patients, the major affected muscles were the deltoids and biceps. Six patients (60%) recovered motor power completely, but the remaining four patients had some degree of paresis. In general, most patients completely regain motor strength after conservative management, but some patients do not recover completely and may have permanent motor weakness. Neurosurgeons should therefore be able to inform their patients of the potential outcomes of NA.

Sensory loss is the third symptom of NA, but sensory changes do not always occur[15]. Sensory changes are generally variable, patchy, and multifocal in distribution and do not always correspond to the site of nerve involvement[15]. Hypesthesia is the most common sensory symptom, followed by a combination of paraesthesia and hypesthesia, and then paraesthesia only[22]. Sensory loss is experienced in the shoulder (most common), arm, forearm, hand, fingers, and/or neck[22]. Sensory symptoms may occasionally occur without motor weakness[18]. In our study, four patients did not have sensory changes (40%) while three other patients had sensory changes with a patchy distribution. We therefore suggest that even if a patient does not have sensory changes, NA should not be excluded, while if a patient has patchily distributed sensory changes, NA should be strongly suspected.

Differential diagnoses of NA include neurologic diseases such as cervical disc herniation, mono- or polyneuritis, spinal cord tumors or pancoast tumors, and orthopedic problems such as rotator cuff injuries or acute calcific tendinitis[15,21]. It is especially difficult to distinguish NA from cervical radiculopathy in the acute phase of NA[14]. We initially misdiagnosed six patients (60%) and cervical spine MRI was performed for four patients. Fortunately, we did not perform any surgical procedures and waited for 3 weeks to confirm our initial diagnoses with neurophysiologic studies.

Neurophysiologic studies are the best modality to confirm a diagnosis of NA. Neurophysiologic findings of NA include abnormal sensory potentials, lack of paraspinal denervation potentials, and abnormal conduction velocities[6,12]. Nerve conduction studies and electromyography (EMG) findings in patients with clinically severe plexopathy reveal absent SNAP, reduced CMAPs, and prolonged F responses, all of which are associated with severe neuropathic changes on EMG and a poorer prognosis for recovery[10]. In general, neurophysiologic studies should be performed 2-3 weeks after the onset of symptoms because there is no degenerative action potential in the acute stage[10]. Therefore, early diagnosis of peripheral neuropathy using neurophysiologic studies is not feasible. We performed neurophysiologic studies in all patients 3 weeks after their initial examination and all of the patients had neurophysiologic findings consistent with NA. We believe that neurophysiologic studies should be taken at least 2 weeks after the development of symptoms in order to prevent false negative results. If the results of neurophysiologic studies are obscure despite the compatibility of the clinical presentations with NA, we recommend a re-examination of neurophysiologic studies after 1-2 weeks.

MRI is a useful tool to diagnoses NA. Magnetic resonance

images may show a diffusely increased T2-weighted signal without a T1-weighted signal change in denervated muscles such as the supraspinatus, infraspinatus, subscapularis, teres minor, deltoid, latissimus dorsi, pectoralis, and rhomboid muscles due to edema. Furthermore, MRI images may show decreased muscular mass and an increased, linear intramuscular T1-weighted signal due to atrophy[8,17]. MRI can also help to exclude rotator cuff injuries and impingement syndrome[8]. However, MRI is insensitive to denervation and less helpful in the diagnosis of NA within 1 month of denervation[4,5].

MRN was first used to diagnose idiopathic brachial plexitis in 2005[16]. MRN is the most sensitive modality to diagnose NA in the acute stage, because MRN can detect the pathology of the peripheral nervous system and allow earlier and more accurate diagnosis[1]. Imaging sequences of MRN include a coronal T1-weighted conventional spin echo sequence, as well as axial and STIR sequences. In patients with NA, STIR imaging can reveal thickening of the involved trunk of the brachial plexus and have a high signal intensity[5,16]. We performed MRN prior to neurophysiologic studies in the last three patients who had typical manifestations of NA. MRN imaging of these three patients revealed thickening and high signal intensity of the involved trunk of the brachial plexus, and we therefore diagnosed acute phase NA and begun appropriate treatment immediately. MRN is therefore an essential diagnostic modality for NA, especially in acute phase NA, and may prevent unnecessary diagnostic procedures or surgical interventions.

The main treatment principle for NA is supportive management, including analgesics and physiotherapy; no specific treatment has been proven to reduce neurologic impairment or improve prognosis[6,15]. Physiotherapy helps patients to recognize and prevent pain and can prevent secondary complications such as shoulder contracture[15,22]. We performed physiotherapy in the early stages to improve the motor weakness of all patients. Use of corticosteroids reduces the time until the start of paresis recovery but does not improve long-term outcome[19,22]. Most patients will generally have good recovery after 2-3 years with supportive management[3,19,20]. We managed all patients conservatively by initiating early physiotherapy, analgesics, and/or steroids, and the pain and motor weakness of all 10 patients improved significantly.

This study has some limitations. First, it is a retrospective study and only a small number of patients were evaluated. Furthermore, MRN was performed in three patients only. However, knowledge of the clinical manifestations and neuroradiologic and neurophysiologic findings of NA can help neurosurgeons who are unfamiliar with NA to avoid unnecessary diagnostic procedures or surgical interventions.

CONCLUSION

NA is a disorder characterized by acute onset of shoulder and arm pain followed by weakness and sensory loss, but the prognosis is very good with conservative management including analgesics and physiotherapy. In our study, NA was misdiagnosed as other neurologic disorders, such as a cervical disc herniation, in six patients (60%). Because NA is misdiagnosed in many cases, awareness and accurate diagnosis of this disorder can help neurosurgeons to determine the most appropriate treatment. To prevent unnecessary surgical procedures or studies, it is important to distinguish NA from other alternative diseases. Neurophysiologic studies are critical for diagnosis of NA, and MRN is very helpful for early diagnosis.

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