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The clinical manifestations and outcomes of neuralgic amyotrophy

[1]Jung Soo Lee *MD PhD*, [2]Yoon Tae Kim *MD PhD*, [3]Joon Sung Kim *MD PhD*, [3]Bo Young Hong *MD PhD*, [3]Lee Chan Jo *MD*, [3]Seong Hoon Lim *MD PhD*

[1]Department of Rehabilitation Medicine, Uijeongbu St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul; [2]Department of Rehabilitation Medicine, National Traffic Injury Rehabilitation Hospital, College of Medicine, The Catholic University of Korea, Seoul; [3]Department of Rehabilitation Medicine, Suwon St. Vincent's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea.

Abstract

Background & Objective: Although the clinical manifestations and outcomes of neuralgic amyotrophy have been previously described, some controversies remain. Thus, we evaluated clinical manifestations and outcomes of patients with neuralgic amyotrophy. *Methods:* We evaluated the clinical and electrodiagnostic data, and the outcomes, of 32 patients with neuralgic amyotrophy.Of the 32 patients, 26 were followed-up for one year after onset of the disease.*Results:*The initial symptoms were pain (50.0%), pain with weakness (21.9%), other sensory symptoms without weakness (6.3%), and painless weakness or atrophy (21.9%). The commonly involved nerves were the median (75.0%), radial (68.8%), suprascapular (50.0%), ulnar (50.0%), axillary (46.9%), and musculocutaneous (40.6%) nerves. The initial symptoms were not associated with nerve involvement. Of all patients, 59% recovered fully, 16% had residual mild weakness without functional disability, and 6% experienced persistent severe weakness and were unable to return to work. Some patients were not evaluated because they were lost to follow-up.
Conclusions: Painless weakness as an initial symptom of neuralgic amyotrophy may be more common than previously noted. Of all patients, 75% enjoyed favorable outcomes by one year after disease onset. These results will be useful when planning treatment strategies and will deepen our understanding of prognosis of neuralgic amyotrophy.

Key words: Neuralgic amyotrophy, outcome, prognosis, recovery.

INTRODUCTION

Neuralgic amyotrophy is a neurological disorder characterized by sudden pain in the shoulder girdle, followed by weakness.[1] Neither the etiology nor the pathophysiology of idiopathic neuralgic amyotrophy is completely understood. However, several authors have proposed that an immune reaction is the principal cause of the disorder.[2,3] Neuralgic amyotrophy is not commonly encountered, and the clinical characteristics of the condition are heterogeneous in nature. Clinical manifestations, the natural history of the disease, and clinical outcomes have been reported by several authors.[4-6] However, the long-term outcomes and the clinical severity of the condition remain controversial; one recent report claimed that neuralgic amyotrophy patients had poor prognoses and developed clinically severe manifestations of disease.[4-7] Another recent report showed that neuralgic amyotrophy was 30-50-fold more common than previously thought.[8] Thus, additional study is needed. We report here the clinical manifestations, electrodiagnostic findings, and long-term clinical outcomes, of a series of Korean patients with neuralgic amyotrophy.

METHODS

This was a retrospective case study. We reviewed the medical records of 32 neuralgic amyotrophy patients who presented between January 2001 and December 2015 to the Departments of Rehabilitation Medicine of St. Vincent's Hospital and Uijeongbu St. Mary's Hospital, Korea. All subjects were diagnosed with electrophysiological study within one month of symptom onset. The inclusion was based on clinical and electrodiagnostic criteria. The weakness with abrupt onset of unilateral or bilateral upper limbs

Address correspondence to: SeongHoon Lim, M.D., Ph.D., Department of Rehabilitation Medicine, Suwon St. Vincent's Hospital, College of Medicine, The Catholic University of Korea. 93 Jungbu-daero, Paldal-gu, Suwon, Kyoungki-do, 442-723, Republic of Korea. Tel: +82-31-249-7650, E-mail: seonghoon@catholic.ac.kr

and that weakness did not caused by cervical roots, brain, spinal cord, trauma or thoracic outlet syndrome. The disorder of brachial plexus was confirmed by electrodiagnosis.[4] Inclusion criteria involved both clinical and electrodiagnostic manifestations as follows: 1) abrupt weakness of either unilateral or bilateral upper limbs which was not caused by lesions of cervical roots, spinal cord or brain, trauma or thoracic outlet syndrome, and 2) brachial plexopathy confirmed by electrodiagnostic test. The exclusion criteria were: 1) any malignancy, 2) diabetes mellitus, or, 3) any other peripheral nerve disease. The study was approved by the Ethics Committee of the Catholic University of Korea.

Demographic, clinical, and electrodiagnostic data were obtained by review of medical records. Electrodiagnostic data were collected with the aid of a Medelec Synergy platform (Oxford Instruments, UK); both nerve conduction study and needle electromyography (EMG) were performed.

RESULTS

We enrolled 32 patients (age 47.7±13.3 years; 23 males, 9 females). The right brachial plexus was involved in 17 patients, the left brachial plexus in 13, and both plexuses in two. The initial symptoms were pain (50.0% of patients), pain with weakness (21.9%), paresthesia or dysesthesia without weakness (6.3%), and painless weakness or atrophy (21.9%).(Table 1) Precipitating events of neuralgic amyotrophy were; common cold (5), herpes zoster infection (1), streptococcal infection (1). The remaining 25 subjects did not recall any particular precipitating event at the time of electrodiagnostic test.

Four of the patients underwent magnetic resonance imaging (MRI) scan. The MRI scan of one patient revealed increased signal intensity of supraspinatus and infraspinatus muscles as a result of denervation.(Figure 1)

The electrodiagnostic findings on the brachial plexus were heterogeneous (Tables 2, 3). As shown, the commonly affected nerves were: median (75.0%), radial (68.8%), suprascapular (50.0%), ulnar (50.0%), axillary (46.9%), and musculocutaneous (40.6%) nerves.(Table 2) The initial symptoms may not correspond with the specific nerve involved.(Table 3)

A total of 26 patients underwent clinical examinations one year post-onset; with six patients (18.7%) being lost to follow-up. Of the 26 patients, 19 (59.4%) recovered fully, five (15.6%) had residual weakness but without functional disability, and two (6.3%) had persistent weakness and were unable to return to work.(Table 4)

DISCUSSION

The reported incidence and outcomes of neuralgic amyotrophy have differed[4,5,9], perhaps because of phenotypic variation, the low incidence of disease, and the fact that primary physicians may be unfamiliar with the condition.[6,10] We explored the clinical manifestations, electrodiagnostic findings, and long-term clinical outcomes of the disease. We found that most of the initial symptoms were pain or sensory discomfort (78.1%), or painless weakness (21.9%). The commonly affected nerves were the median, radial, suprascapular, ulnar,

Table 1: Clinical characteristics of patients

Characteristics	No. of patients (%)
Age (years)	47.7±13.3
Male/ Female	23 (71.9) / 9 (28.1)
Age (years)	47.7±13.3
Involved side. Right/Left/Bilateral	17(53.1)/13(40.6)/2(6.3)
Initial presentation(symptoms)	
Pain	16 (50.0)
Pain and weakness	7 (21.9)
Paresthesia and dysthesia	2 (6.3)
Painless weakness and atrophy	7 (21.9)
Precipitating event	
Non-specific to recall	25 (78.1)
Common cold	5 (15.6)
Herpes zoster infection	1 (3.1)
Streptococcus agalactiae infection	1 (3.1)

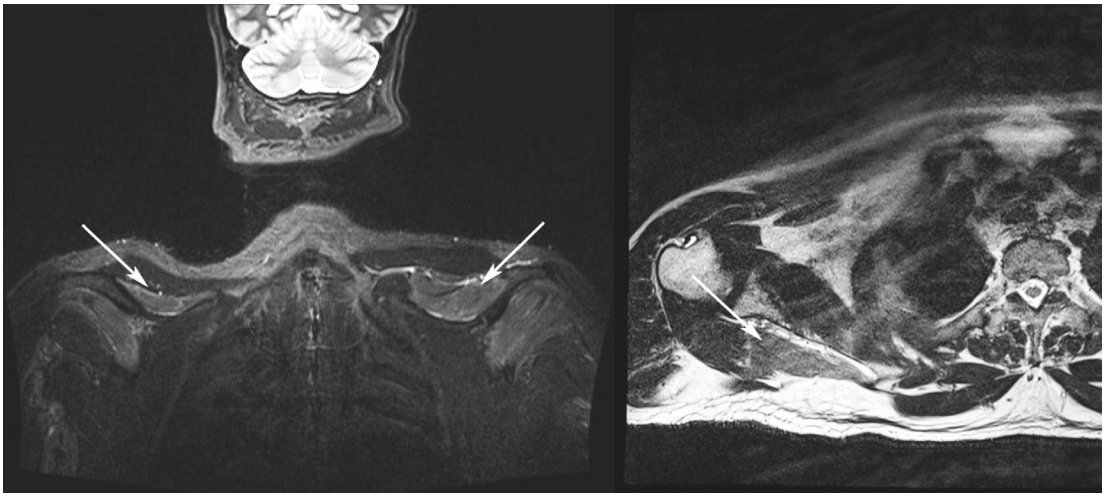

Figure 1: MRI images of a patient with bilateral neuralgic amyotrophy. T2 scan demonstrating increased signal intensity of supraspinatus muscles as a result of denervation. Left showing denervation of bilateral supraspinatus muscles in coronal plane, and right showing denervation of right supraspinatus muscle in transverse plane. The arrows indicatesdenervated supraspinatus muscles.

axillary, and musculocutaneous nerves. At one year after disease onset, almost 75% of patients had favorable outcomes, but 6% of the patients exhibited persistent weakness of the affected nervesthat affected the patients functionally.

Our study was retrospective in nature. Thus, we focused on clinical presentations and outcomes. We found that painless weakness was the first symptom in 21.9% of the patients. This incidence is higher than that of several previous reports but lower than that of one such report.[4,5,11] These differences can be partly explained by phenotypic variation, a concept introduced by van Alfen. However, the current state of knowledge on neuralgic amyotrophy remains inadequate. Taking our data and previous results together, it appears that pain is usually the initial symptom, but painless weakness or atrophy is a significant initial symptom in 7-30% of patients.[5,11]

Table 2: Frequency of the nerve affectation and percent over 32 patients

Nerve	Number of lesions	% over 32 patients
Motor Nerves		
Spinal accesory	1	3.1
Suprascapular	16	50.0
Dorsal scapular	5	15.6
Long thoracic	3	9.4
Lateral pectoral	1	3.1
Thoracodorsal	1	3.1
Axillary	15	46.9
Musculocutaneous	13	40.6
Radial	22	68.8
Median	24	75.0
Ulnar	16	50.0
Sensory Nerves		
MACN[1]	4	12.5
LACN[2]	1	3.1
Radial	1	3.1
Median	3	9.4
Ulnar	3	9.4

1. MACN: Medial antebrachial cutaneous nerve, 2. LACN: Lateral antebrachial cutaneous nerve.

Table 3: Initial symptoms and involved nerves for each subjects

No.	Initial Symptom	Involved nerves
1	Paresthesia at whole upper limb	Radial, median, ulnar
2	Painless weakness	Spinal acessory, axillary, musculocutaneous, radial, median
3	Pain at shoulder	Median, ulnar
4	Pain at shoulder and arm	Median, ulnar
5	Pain at shoulder	Axillary, suprascapular
6	Painless weakness	Dorsal scapular, suprascapular, axillary, radial, median, ulnar
7	Painless weakness	Dorsal rami (C6-T1), radial, median, ulnar, radial
8	Pain at whole upper limb	Axillary, radial, median
9	Pain at shoulder	Long thoracic, suprascapular, radial, median, ulnar
10	Pain at neck	Radial, median, ulnar
11	Pain at shoulder and elbow	Musculocutaneous, radial, median, ulnar
12	Pain at shoulder	Suprascapular, axillary, musculocutaneous
13	Pain at arm and wrist	Dorsal rami (C7), Radial, median, ulnar
14	Pain at shoulder	Dorsal rami(C4-7), lateral pectoral, suprascapular, axillary, radial, median, ulnar
15	Painless weakness	Dorsal scapula, long thoracic
16	Pain at shoulder	Radial, median
17	Pain at shoulder	Dorsal rami (C5-T1), suprascapular, musculocutaneous, radial, median, ulnar
18	Pain at shoulder	Suprascapular, musculocutaneous, radial, median
19	Paresthesia at forearm and hand	Suprascapular, musculocutaneous, radial, median, ulnar
20	Pain at shoulder	Dorsal scapular, suprascapular
21	Painless weakness	Dorsal rami (C5-T1), dorsal scapular, suprascapular, axillary, musculocutaneous, radial, median
22	Pain at shoulder	Radial, median, ulnar
23	Pain and weakness at shoulder	Radial, median, ulnar
24	Painless weakness at both upperlimb	Bilateral suprascapular, long thoracic, thoracodorsal, axillary, radial, median, ulnar,
25	Painless weakness at both upperlimb	Bilateral suprascapular, musculocutaneous, left axillary
26	Pain, weakness and atrophy at shoulder	Suprascapular, axillary, musculocutaneous, radial, median
27	Pain at shoulder	Suprascapular, axillary, musculocutaneous
28	Pain and weakness at shoulder	Suprascapular, axillary, musculocutaneous
29	Pain and weakness at shoulder	Axillary, musculocutaneous
30	Pain and weakness at shoulder	Suprascapular, axillary, musculocutaneous, median
31	Pain and weakness at shoulder	Dorsal scapular, radial, median
32	Pain and weakness at shoulder and arm	Radial, median, ulnar

Cruz –Mrtinez reported that over 90% of affected nerves recovered by two years after disease onset.[4] However, van Alfen reported that two-thirds of their patients experienced persistent pain and paresis.[5] Tsairisstated that nearly 80% of all patients recovered normal function.[11] We found that nearly 75% of our patients recovered, without any residual functional disability, within one year of disease onset. Thus, favorable outcomes may be expected in 33-80% of patients. Given such among-study heterogeneity, further study focusing on functional recovery is urgently needed.

Table 4: Clinical outcome at one year after onset

Recovery without sequele	19(59.4%)
Recovered with mild weakness (Able to work)	5 (15.6%)
Recovered with definite weakness (Unable to work)	2 (6.3%)
Lost to follow up	6 (18.7%)

The most affected nerves also varied among studies. We found that the median, radial, suprascapular, ulnar, axillary, and musculocutaneous nerves were commonly involved. A previous report showed that upper trunk nerve involvement was common; however, we did not find such selectivity of involvement.[4]

Regardless of the initial symptoms, the entire brachial plexus, from the roots to the peripheral nerves, may be involved. Therefore, clinicians and specialists in electrodiagnostic medicine should carefully evaluate the entire brachial plexus if neuralgic amyotrophy is suspected.There has been anecdotal evidence that corticosteroids have a favorable effect on pain and recovery.[12] There is need for proper clinical randomized trial to evaluate the effectiveness of the various therapeutic intervention for functional recovery in neuralgic amyotrophy. One randomized controlled trial comparing prednisolone to placebois currently awaiting formal reporting.[12]

Our study had several limitations. First, we had no follow-up data obtained later than one year after disease onset; long-term outcomes were thus not addressed. However, patients who experience marked improvements may decide not to keep hospital appointments. We thus decided to evaluate the 1-year follow-up data to exclude selection bias. Second, our study was retrospective in nature; we cannot contribute to the recent debate on disease incidence. Only a small proportion of the patients revealed some precipitating factors. It may be due to recall bias or limitation of retrospective study design. Third, our number of subjects was small, because the disease is relatively rare. However, we excluded diabetes, since diabetes can greatly influence the results of electrodiagnostictest.[13] This enabled us to have more exact electrodiagnostic data.

In conclusion, painless weakness as an initial symptom of neuralgic amyotrophy may be more common than previously thought. Of all patients, 70% experienced favorable outcomes by 1 year after disease onset.

DISCLOSURES

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Conflict of Interest: None

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