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# A Clinical Study of Benign Focal Amyotrophy

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*We reviewed the clinical, electrophysiological, radiological, and histopathological findings in 25 patients with benign focal amyotrophy.*
*There were 14 patients with upper limb type and 11 with lower limb type. 18 patients had unilateral involvement and 7 had bilateral involvement asymmetrically. The characteristic clinical features were sporadic occurrence, predominance in young males, nonprogressive course or initial progression for 1 to 3 years followed by stationary state, segmental distribution of muscle weakness and atrophy localized to one limb or both homologous limbs markedly asymmetrically, and absence of any definite sensory loss or central nervous system involvement. The electrophysiological, radiological, and muscle histopathological findings suggested chronic focal anterior horn cell disease.*
*Although the prevalence of this disease is still unknown, the importance of recognition is being emphasized because of its common occurrence in our country and the benign prognosis.*

**Key Words** : *Benign focal amyotrophy, Anterior horn cell disease.*

## INTRODUCTION

Benign focal amyotrophy has been recognized as a limited form of anterior horn cell disease with a benign prognosis. From the first description of "juvenile muscular atrophy of unilateral upper extremity" by Hirayama et al in 1959, about 200 similar cases have been reported mainly from India and Japan. But only a few cases have been reported from western countries. This syndrome has been described under various names such as juvenile muscular atrophy of unilateral upper extremity (Hirayama et al., 1959 ; Singh et al., 1980), benign focal amyotrophy (Adornato et al., 1978), wasted leg syndrome (Prabhakar et al., 1981), and monomelic amyotrophy (Gourie-Devi et al., 1984).

This disease is characterized by sporadic occurrence in young age, male predominance, insidious onset with progression for several years followed by a stationary state, and muscle wasting and weakness confined to upper or lower extremity unilaterally or markedly asymmetrically. The electrophysiological findings suggested anterior horn cell involvement and a pathological study demonstrated focal lesion in the anterior horn of the spinal cord (Hirayama et al., 1987). Although several cases have been reported in Korea (Choi et al., 1982 ; 1987 ; Cheong et al., 1992), comprehensive studies of this disease are rarely available. We report our experiences of 25 patients.

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## MATERIALS AND METHODS

A total of 25 patients who were diagnosed with benign focal amyotrophy at the Neurology Department of Seoul National University Hospital from March 1984 to March 1993, were analyzed. The criteria for diagnosis were as follows ; (1) muscle weakness and wasting localized to one limb or both homologous limbs markedly asymmetrically, (2) nonprogressive course or initial progression followed by stationary state, (3) absence of any definite sensory loss or central nervous system involvement, (4) absence of the radiological evidence of a compressive lesion of the spinal cord.

Detailed histories were obtained including occupation, vaccination, exposure to toxins, trauma, and viral infections. There was no history suggestive of poliomyelitis or familiar neuromuscular disease. General physical and neurological examinations were carried out on all patients. Routine laboratory findings such as complete blood counts, liver function tests, serum glucose levels, serum muscle enzyme levels, serologic tests for syphilis, and urinalysis were normal in all patients. Cerebrospinal fluid examinations were done in 12 of 25 patients and were normal.

Electromyographic (EMG) studies were done in all patient with a DISA Neuromatic 2000-C electromyograph. Motor and sensory nerve conduction studies were performed in the affected and the unaffected homologous limbs. Motor nerve conduction studies were done in median and ulnar nerves in upper limbs and peroneal and posterior tibial nerves in lower limbs. The terminal latency, the amplitude of compound muscle action potential (CMAP), motor nerve conduction velocity, and F-wave latency were determined. Sensory nerve conduction studies were done in median, ulnar, and sural nerves. The amplitude of compound nerve action potential (CNAP) and sensory nerve conduction velocity were also determined. Needle EMG studies were performed in the affected, clinically unaffected homologous limb, and other limbs. Somatosensory evoked potential studies were performed in 15 patients.

Spine CT scans or metrizamide CT myelograms were taken in 8 patients and spine MR imaging was done in 15 patients.

Muscle biopsies were taken from the affected muscles or clinically unaffected neighbouring muscles in 13 of 25 patients. Sural nerve biopsies were performed in 2 patients. Routine and special histopathological procedures including muscle histochemistry and special stainings were done. For sural nerve biopsy, electromicroscopic examination was also performed together with routine histology.

## RESULTS

During nine years from March 1984 to March 1993, 189 adult patients with chronic anterior horn cell diseases were admitted to the Neurology Department of Seoul National University Hospital. Amyotrophic lateral sclerosis formed the largest group (144 cases, 76.2%), followed by benign focal amyotrophy (25 cases, 13.2%) and spinal muscular atrophy (20 cases, 10.6%).

Among 25 patients with benign focal amyotrophy, 14 had upper limb involvement (Group U) and 11 had lower limb involvement (Group L). The age of onset ranged from 16 to 36 years with a mean of 23.6 years. In group U, the mean age of onset was

**Table 1.** Sex, Age, and Affected Limbs in 23 Patients

| | Gr. U (N=12) | Gr. L (N=11) | Total (N=23) |
|---|---|---|---|
| Sex (No.of pts) | | | |
| Male | 11 | 8 | 19 |
| Female | 1 | 3 | 4 |
| Age of onset (yr) | | | |
| Mean | 20.3 | 28.5 | 24.2 |
| Range | 17-33 | 17-36 | 17-36 |
| Affected limb (No.of pts) | | | |
| Unilateral Rt | 7 | 5 | 12 |
| Lt | 1 | 3 | 4 |
| Bilateral Rt>Lt | 1 | 1 | 2 |
| Rt<Lt | 3 | 2 | 5 |

19.8 years (16 to 33 years), which was considerably younger than group L with a mean age of 28.5 years (17 to 36 years). There were 21 males and only 4 females. 18 patients had unilateral limb involvement and 7 had asymmetrical bilateral involvement (Table 1).

### Clinical manifestations

Upper limb involvement group (Group U): Among 14 patients with upper limb involvement, 10 patients (71.4%) had unilateral involvement and 4 (28.6%) had bilateral involvement. In 10 patients with unilateral involvement, the right side was affected in 7 and the left in 3 (Table 1). In the majority of the patients (13/14 cases), the chief complaint was difficulty in hand gripping and fine hand movements. In those 13 patients, the atrophic and weak muscles were invariably confined to intrinsic hand muscles, flexors and extensors of the forearm. Biceps brachi and deltoid muscles were rarely involved (Fig. 1). One patient showed severe muscle wasting and weakness in proximal muscles such as deltoid, sternocleidomastoid, trapezius, supraspinatus, and pectoralis major (Fig. 2). In this patient, intrinsic hand muscles and forearm muscles were intact, although arm muscles showed minimal muscle wasting in biceps and triceps brachi. The muscle wasting and weakness had not been progressive since the time of detection in 4 patients. But it was progressive for 8 months to 3 years, and then stationary in the remaining 10 patients. Fine, irregular, tremulous movements involving the fingers were noted in 8 of 14 patients (57.1%). The tremor was present in resting state and aggravated by stress and voluntary actions. Fasciculation was reported in 5 patients, mainly in the forearm. Four patients noted stiffness of hands aggravated by cold exposure. Feelings of numbness were complained of by 4 patients but objective sensory loss could not be detected. In one patient, clawing of the fingers was noted. Deep tendon reflexes in the affected upper limbs were hyporeactive in 6 patients (42.8%), normoreactive in 6 (42.8%), and hyperreactive in 2 (14.3%) (Table 2).

Lower limb involvement group (Group L): The patients in group L were incidentally detected by muscle atrophy or limping gait. There was unilateral involvement in 8 (72.7%) and bilateral in 3 (27.3%).

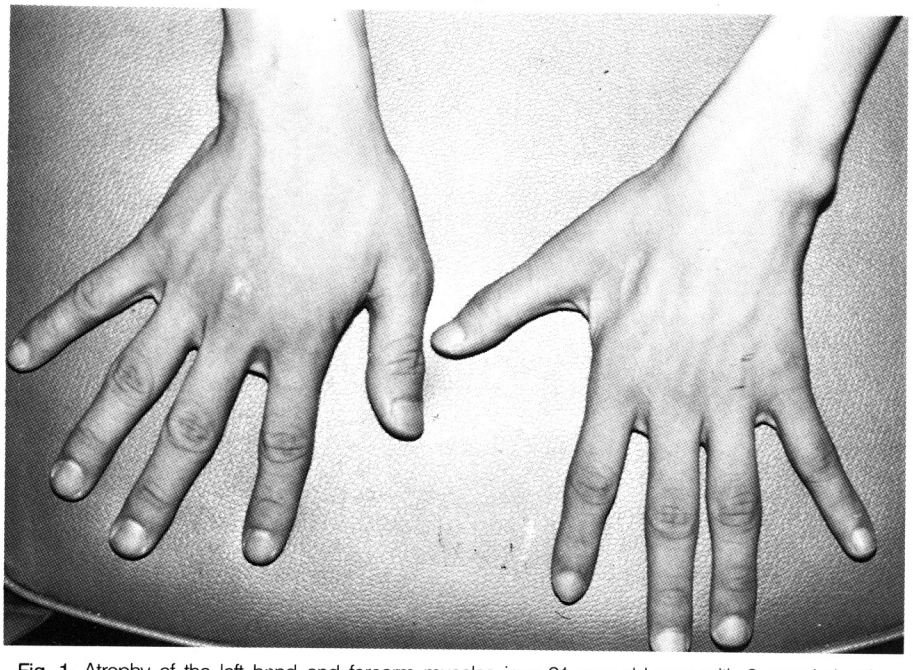

Fig. 1. Atrophy of the left hand and forearm muscles in a 21 year-old man with 3 years' duration.

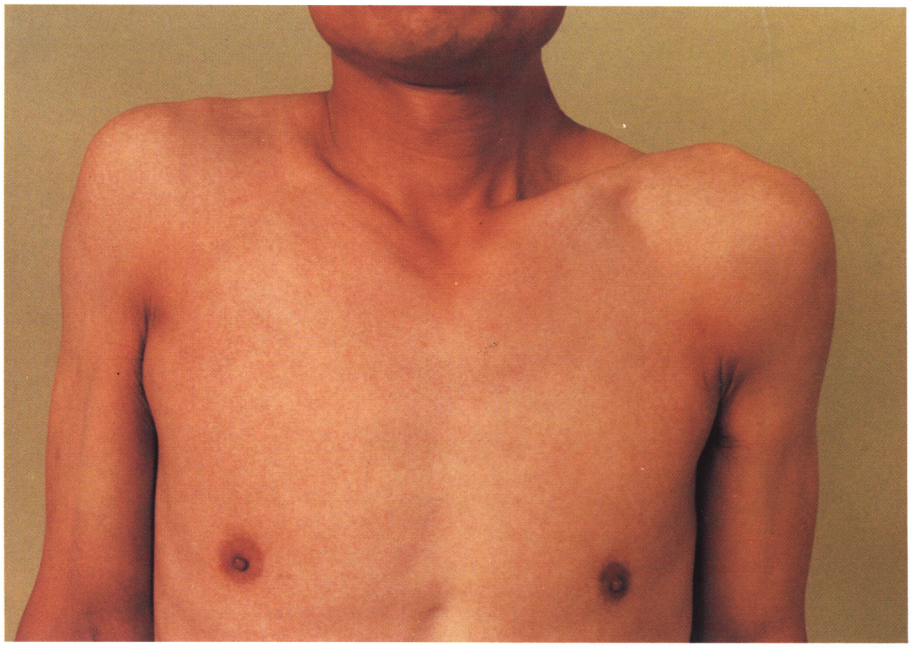

Fig. 2. Atrophy of the left shoulder girdle and proximal arm muscles in a 30 year-old man with 9 years' duration.

Table 2. Clinical Manifestations in 23 Patients

| Clinical Manifestations | Patient No.(%) | | |
|---|---|---|---|
| | Gr. U (N=12) | Gr. L (N=11) | Total (N=23) |
| Weakness/wasting | 12 (100.0%) | 11 (100.0%) | 23 (100.0%) |
| Tremor | 7 (58.3%) | 0 (0.0%) | 7 (30.4%) |
| Fasciculation | 5 (41.7%) | 3 (27.3%) | 8 (34.8%) |
| Cold paresis | 4 (33.3%) | 1 (9.1%) | 5 (21.7%) |
| Numbness | 4 (33.3%) | 3 (27.3%) | 7 (30.4%) |
| Flexion deformity | 1 (8.3%) | 5 (45.5%) | 6 (26.1%) |
| DTR | | | |
| Hyporeactive | 6 (50.0%) | 6 (54.5%) | 12 (52.2%) |
| Normoreactive | 4 (33.3%) | 4 (36.4%) | 8 (34.8%) |
| Hyperreactive | 2 (16.7%) | 1 (9.1%) | 3 (13.0%) |

In patients with unilateral involvement, the right side was affected in 5 and the left in 3 (Table 1). The affected muscles consisted of the thigh muscles in one patient (9.1%), crural muscles in 3 (27.3%), and both thigh and crural muscles in 7 (63.6%) (Fig. 3). One patient showed asymmetrical distribution of muscle atrophy in the right thigh and the left crural muscles. The clinical course was nonprogressive in 6 patients (54.5%) and initial progression for 1 to 2 years followed by a stationary state in 5 (45.5%). Muscle fasciculation was noted in 3 patients (27.3%). Three patients (27.3%) complained of sensory numbness and one (9.1%) reported stiffness of the involved leg on exposure to cold. Pes cavus deformity was observed in 5 (45.5%) and hammer toe deformity was also accompanied in 2 of those patients. Deep tendon reflexes were hyporeactive in 6 (54.5%), normoreactive in 4 (36.4%), and hyper-

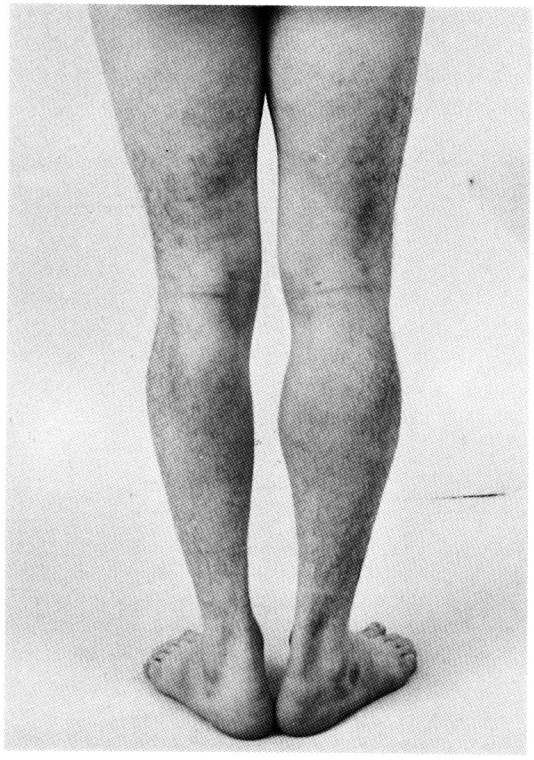

**Fig. 3.** Atrophy of the crural muscles of the left leg in a 31 year-old man with 2 years' duration.

reactive in 1 (9.1%). Long tract signs were absent in all patients (Table 2).

### Electrophysiological findings

Motor nerve conduction studies in the affected limbs were normal in 14 of 25 patients (56.0%) (Table 3). Reductions in CMAP amplitudes were noted in 10 patients (40.0%), prolonged terminal latencies in 6 (24.0%), and prolonged F-wave latencies in 7 (28.0%). Motor nerve conduction velocities were slowed in 4 patients (16.0%), but never fell below 70% of the lower limit of normal data in our EMG Laboratory. The slowing of motor nerve conduction velocity was accompanied by the reduction of CMAP amplitudes and seemed to be associated with the degree of atrophy of the muscle. Conduction block phenomenon was not observed and motor nerve conduction studies in the unaffected homologous limbs were normal in all patients. Sensory nerve conduction studies were quite normal in all patients.

Needle EMG studies in the affected muscles invariably showed evidence of denervation such as fibrillation potentials, positive sharp waves, giant motor unit potentials, and reduced interference patterns. The denervation in EMG was not observed in clinically unaffected remote muscles of the same limb. In 14 of 25 patients, the clinically unaffected contralateral limb was also examined and mild denervation was noted in 6 of 14 patients. The lower limb was studied in 3 cases of group U and the upper limb in 5 of group L, which were all normal.

The somatosensory evoked potentials for median or posterior tibial nerves were normal in 8 patients tested in group U, and in 7 tested in group L.

### Radiological findings

Spine CT scans or metrizamide CT myelograms were taken in 8 patients, and spine MR imaging was done in 15 patients. There was no evidence of cord compression and no definite foraminal or osseous abnormality. In one patient with atrophy of the left hand and forearm muscles, cervical MR imaging revealed segmental atrophy in the lower cervical cord, especially in the anterior horn region, which correlated with clinical and electromyographic

**Table 3.** Results of Motor Nerve Conduction Studies in 23 Patients

| Findings | Gr. U (N=11) | Gr. L (N=11) | Total (N=23) |
|---|---|---|---|
| Normal | 6 | 8 | 14 (60.9%) |
| Abnormal | | | |
| Low amp. CMAP | 6 | 3 | 9 (39.1%) |
| Prolonged T.L. | 2 | 3 | 5 (21.7%) |
| Slow NCV | 1 | 2 | 3 (13.0%) |
| Prolonged F.L. | 3 | 3 | 6 (26.1%) |

T.L. ; terminal latency, F.L. ; F-wave latency

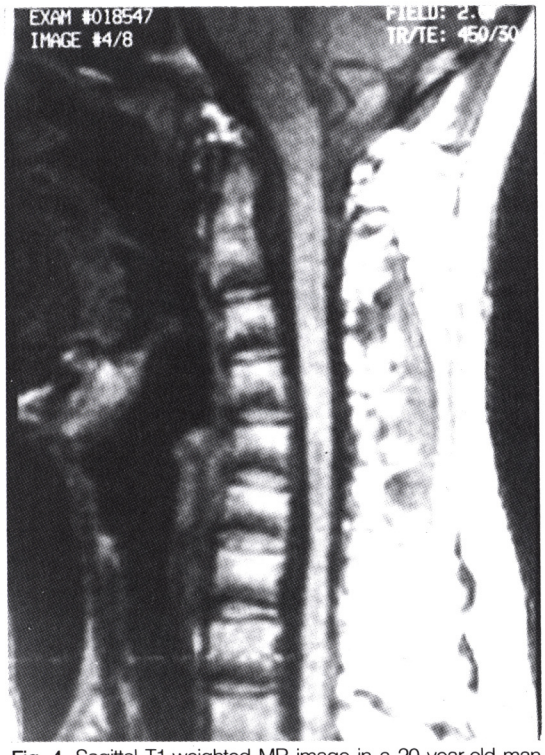


Fig. 4. Sagittal T1-weighted MR image in a 20 year-old man with hand and forearm atrophy of 1 year's duration showing segmental atrophy in the lower cervical cord.

findings (Fig. 4). Lumbar spine MR was performed in 8 out of 11 patients of group L, but showed no diagnostic abnormalities. In two patients with unilateral upper limb involvement, plain X-ray of cervical spine was only taken. These two patients showed typical clinical and electrophysiological characteristics of group U, and were included in this study.

### Histopathological findings

The results of muscle biopsies in 13 patients were quite variable from normal to neurogenic atrophy or myopathic change according to the muscle selected In 8 of 13 patients (61.5%), histopathological findings from clinically and electrophysiologically unaffected muscles (deltoid in 4, biceps brachi in 1, brachioradialis in 2, and vastus lateralis in 1) revealed no diagnostic abnormalities. In 4 patients (30.8%) whose muscle biopsies were taken from the clinically affected muscles, scattered small angulated myofibers and grouped atrophy were observed, suggestive of neurogenic atrophy (Fig. 5). In one patient (7.7%), muscle biopsy revealed marked atrophic myofibers, giant cell transformation of myofibers with nuclear clumping, and diffuse fatty infiltration, which were suggestive of end stage muscle disease.

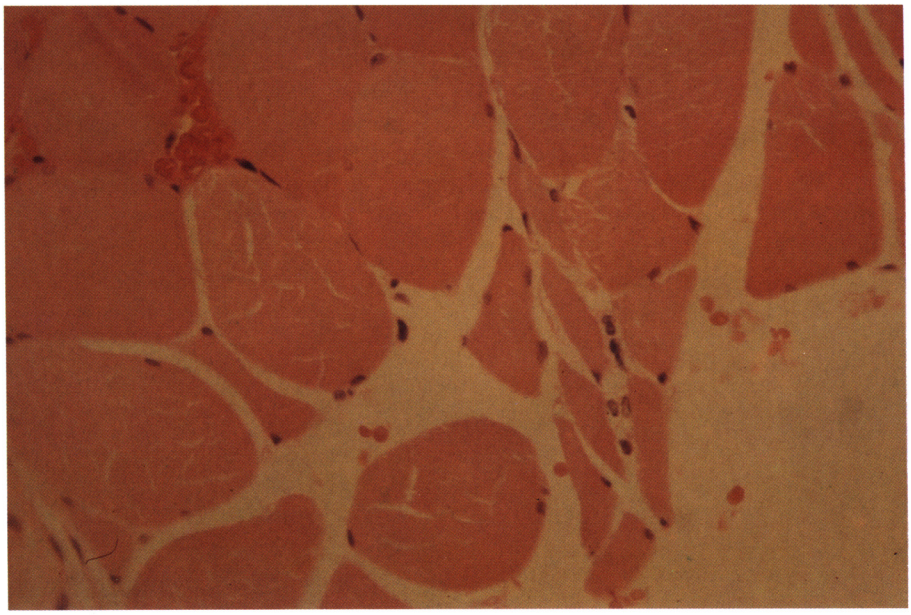

Fig. 5. Neurogenic atrophy with grouping of small angulated atrophic myofibers (H & E stain).

In two patients with lower limb involvement and pes cavus deformity, sural nerve biopsies were performed and proved to be normal.

## DISCUSSION

"Benign focal amyotrophy" was first described by Adornato et al as a limited form of lower motor neuron disease confined to the upper extremities unilaterally or markedly asymmetrically (Adornato et al., 1978). Although this term emphasized the benign nature of the disease in contrast to fatal amyotrophic lateral sclerosis, a similar description had already been made by Hirayama et al in 1959 and 1963 under the name of "juvenile muscular atrophy of unilateral upper extremity". In the first series by Hirayama et al in 1959, the authors reported twenty young male patients with unilateral hand and forearm muscle involvement, but subsequent reports included cases with bilateral involvement also (Hashimoto et al., 1978). In 1981, Prabhaker et al reported 40 cases with nonprogressive muscle atrophy of unilateral lower extremity and called it "wasted leg syndrome". The term "monomelic amyotrophy" was introduced in 1984 to emphasize the restrictive nature of muscle atrophy and weakness of one limb, either upper or lower extremity (Gouri-Devi et al., 1984 ; 1986). But it might be inappropriate because there are many cases with bilateral involvement.

The major clinical features in our series were not different from previous reports ; muscle weakness and wasting localized to one limb or both homologous limbs markedly asymmetrically ; nonprogressive course or initial progression for 1 to 3 years followed by stationary state ; predominance in young males ; sporadic occurrence ; absence of any definite sensory loss or central nervous system involvement. The mean age of onset in group U was 19.8 years, which was significantly younger than group L (28.5 years). The discrepancy in the age of onset between group U and group L has been described in previous reports (Gouri-Devi et al., 1984), and raised a possibility that these two groups might be of a different origin. In fact, many Japanese researchers have not used the term "benign focal amyotrophy" and they have regarded the lower limb type as a different disease entity.

The typical pattern of muscle atrophy limited to the hand and forearm, especially in the ulnar side, was prominent in cases of group U, except for one patient with proximal involvement. Cases with atrophy of the shoulder girdle and proximal upper arm have already been described under the diagnosis of benign focal amyotrophy or monomelic amyotrophy in previous reports (Gouri-Devi et al., 1984 ; Metcalf et al., 1987). Although Hirayama's series emphasized the sparing of the brachioradialis muscle, our studies of the brachioradialis muscle showed electromyographic denervation in 3 of 4 cases tested and neurogenic atrophy in 2 of 4 biopsied specimens. Unlike the relatively stereotypic and focal distribution of atrophy in group U, the lower limb group showed a diffuse involvement of thigh and crural muscles in 7 of 11 cases (63.3%) as in Prabhaker's series (65%).

The coarse and irregular tremor of the fingers seen in 8 patients (57.1%) of group U is a well-known finding in this disease, but has also been described in Kugelberg-Welander disease and childhood spinal muscular atrophy and termed "benign fasciculation" or "minipolymyoclonus" (Byers et al., 1961 ; Spiro, 1970). These tremulous movements in patients with muscle atrophy have been considered useful in differentiating neuropathies from myopathies (Spiro, 1970). "Cold paresis" in 4 patients of group U and 1 patient of group L indicated aggravation of weakness and muscle stiffness by cold. This phenomenon was originally described by Hirayama et al., and thought to be caused by autonomic disturbance or increased sensitivity of atrophic muscles to cold (Hirayama et al., 1963 ; Gouri-Devi et al., 1984). Increased plantar reflex in the affected limbs seen in 3 patients (12.0%) in our series, has previously been reported (Gouri-Devi et al., 1984 ; Singh et al., 1980), and might suggest possible involvement of the upper motor neuron. But this was not thought likely because there were no additional signs of upper motor neuron involvement.

Electomyographic evidence of denervation and relatively normal nerve conduction studies in the present and other reported series were consistent with a chronic anterior horn cell disease. There was no conduction block phenomenon suggestive of focal motor neuropathy. The denervation pattern in EMG was confined to the muscles which were innervated by a few segmental levels of the spinal cord and accounted for the focal nature of the disease. Absence of widespread denervation was an important EMG finding for exclusion of fatal amyotrophic lateral sclerosis and spinal muscular

atrophy. In our 14 patients out of 18 with unilateral involvement, the clinically unaffected contralateral limb was also studied, that revealed a mild denervation pattern in 6 patients. This finding was indicative of the relatively common occurrence of bilateral involvement in this disease, either clinically or subclinically. A distal variety of chronic spinal muscular atrophy has been recognized in several reports (Meadows et al., 1969 ; O'Sullivan et al., 1978 ; Riggs et al., 1984), which had the clinical features of muscle wasting and weakness virtually mimicking benign focal amyotrophy with bilateral involvement. The slowly progressive course and the presence of electromyographic denervation in both upper and lower limbs could distinguish the distal type of chronic spinal muscular atrophy from benign focal amyotrophy. Riggs et al suggested that benign focal amyotrophy might be a variant of chronic spinal muscular atrophy that remained clinically focal for decades or perhaps indefinitely (Riggs et al., 1984).

Spine MR imaging or metrizamide spine CT myelograms should be performed to exclude the correctable conditions such as spondylosis, herniated disc, cord tumor, syringomyelia, or arteriovenous malformation, etc. We experienced a case of a 40-year-old male with focal atrophy of the right upper limb, in whom the cervical spine MR imaging revealed an intradural extramedullary tumor at $C_4$-$C_8$ level, and the histological diagnosis of the tumor was meningioma. MR and metrizamide CT studies of the cervical cord in cases of upper limb type have been reported to show focal and segmental atrophy in the lower cervical cord limited to the anterior horn region and have provided useful information to confirm the diagnosis (Metcalf et al., 1987 ; Biondi et al., 1989 ; Hirabuki et al., 1991). In one patient of our series, MR imaging revealed segmental atrophy in the lower cervical cord, especially in the anterior horn region, which correlated with clinical and electromyographic findings (Fig. 4). We could not observe cord atrophy in the cases in group L, as in other studies (Cheong et al., 1992).

The histopathological muscle findings showed a wide variation by biopsy site. In 8 cases (61.5%), muscle biopsies from clinically and electrophysiologically unaffected muscles of the involved or uninvolved homologous limbs revealed no diagnostic abnormalities. Normal muscle biopsy findings were reported in 7.2% to 18.4% even in cases of amyotrophic lateral sclerosis (Achari et al., 1974 ; Mumenthaler, 1969). The common occurrence of normal biopsy findings in our study was thought to suggest the focal nature of this disease. When atrophic muscles were selected, the biopsy showed neurogenic change in 4 cases and myopathic change in 1 case. The myopathic change seen in our series might not be of genuine myopathy. Myopathic change in chronically denervated muscles has been well documented in previous reports and regarded as nonspecific changes during the chronic denervation process (Drachman et al., 1967 ; Achari et al., 1974).

Although the clinical, electrophysiological, radiological, and muscle histopathological findings suggested segmental anterior horn lesions in this disease, necropsy findings have been reported from only two patients with upper limb involvement (Hirayama et al., 1987 ; Araki et al., 1989). The necropsy study showed lesions only in the anterior horns of the spinal cord at C5-T1, consisting of shrinkage and necrosis, degeneration of large and small nerve cells, and mild gliosis (Hirayama et al., 1987).

The pathogenetic mechanism underlying this disease remains uncertain. There has been no evidence of trauma, toxins, nutritional factors, or viral and syphilitic infections as possible etiological factors. Circulatory insufficiency in the territory of the spinal cord has been suggested (Hirayama et al., 1987), and Breig et al postulated that forward bending of the neck would flatten the cord from front to back and stretch the vessels ; this in turn could induce a secondary vascular insufficiency (Breig et al., 1966). Several reports have recently proposed "tight dural canal in flexion" and suggested that anterior shift of the posterior lower cervical dura mater during neck anteflexion would compress the cervical spinal cord and would play an important role in pathogenesis (Kitagawa et al., 1992 ; Murata et al., 1992 ; Tokumaru et al., 1989). There has been no proposed pathogenetic mechanism in cases of lower limb type, but the fact that the lumbar spinal cord is very vulnerable in systemic circulatory failure suggests the possibility of vascular insufficiency in lower limb type. The striking features of predilection in young males and geographical distribution mainly in eastern countries may offer some clues to the unknown pathogenesis of this disease.

Differential diagnoses include classic motor neuron disease such as amyotrophic lateral scler-

osis, juvenile spinal muscular atrophy, late motor neuron degeneration after poliomyelitis, residua of brachial plexopathy, syringomyelia, and compressive lesions of the spinal cord. Amyotrophic lateral sclerosis can be differentiated by the age of onset, progressive clinical course, positive pyramidal tract signs, and widespread involvement in more than three limbs and bulbar muscles. Juvenile spinal muscular atrophy, especially distal type, is characterized by bilaterally symmetric involvement in upper and lower limbs, slowly progressive course, and positive family history in many cases (Meadows et al., 1969 ; O'Sullivan et al., 1978 ; Riggs et al., 1984). Late motor neuron degeneration after poliomyelitis occurs several years after the definite history of acute poliomyelitis and is progressive (Campbell et al., 1969 ; Mulder et al., 1972). Brachial neuritis often shows severe pain, selective sensory involvement, and no fasciculations (Spillane JD, 1943). Structural lesions such as syringomyelia, tumors, arteriovenous malformations, spondylosis, and herniated discs have sensory involvement and characteristic radiological findings.

Benign focal amyotrophy constituted 13.2 % of chronic anterior horn cell diseases admitted to the Neurology Department of Seoul National University Hospital, a major teaching hospital of 1100 beds capacity in Seoul, during a nine years' period. The importance of recognition of this disease is being raised because of its common occurrence in our country and the benign prognosis.

## REFERENCES


Achari AN and Anderson MS : *Myopathic changes in amyotrophic lateral sclerosis : Pathologic analysis of muscle biopsy change in 111 cases. Neurology 24 : 477-481, 1974.*

Araki K, Ueda Y, Michinaka C, Takamasu M, Takino T, Konishi H : *An autopsy case of juvenile muscular atrophy of unilateral upper extremity (Hirayama's disease). Nippon Naika Gakkai Zasshi 78 : 674-675, 1989.*

Adornato BT, Engel WK, Kucera J, Bertorini TE : *Benign focal amyotrophy. Neurology 28 : 399, 1978.*

Biondi A, Dormont D, Weitzner IJ, Bouche P, Chaine P, Bories J : *MR imaging of the cervical cord in juvenile amyotrphy of distal upper extremity. Am J Neuroradiol 10 : 263-268, 1989.*

Breig A, Tumbull I, Hassler O : *Effects of mechanical stress on the spinal cord in cervical spondylosis. A study on fresh cadaver material. J Neurosurg 25 : 45-54, 1966.*

Campbell AMG, Williams ER, Pearce J : *Late motor neuron degeneration following poliomyelitis. Neurology (Minneap) 19 : 1101-1106, 1969.*

Cheong KH, Cho PZ, Sunwoo IN, Park YK, Lee SA, Kim KW, Park KD : *Clinical characteristics of benign focal amyotrophy. J Korean Neurol Assoc 10 : 447-456, 1992.*

Choi IS : *2 cases of juvenile muscular atrophy confined to upper limb. J Korean Med Assoc 25 : 669-670, 1982.*

Choi IS : *Benign focal amyotrophy. J Korean Med Assoc 30 : 1371-1374, 1987.*

Drachman DB, Murphy SR, Nigam MP, Hills JR : *"Myopathic" changes in chronically denervated muscle. Arch Neurol 26 : 14-24, 1967.*

Gourie-Devi M, Suresh TG, Shankar SK : *Monomelic amyotrophy. Arch Neurol 41 : 388-394, 1984.*

Gouri-Devi M, Suresh TG : *Benign focal amyotrophy or monomelic amyotrophy. Arch Neurol 43 : 1223, 1986.*

Hashimoto O, Asada M, Ohta M : *Clinical observations of juvenile nonprogressive muscular atrophy localized in hand and forearm. J Neurol 211 : 105-110, 1976.*

Hirabuki N, Mitomo M, Miura T, Hashimoto T, Kawai R, Kozuka T : *Computed tomographic myelography characteristics of spinal cord atrophy in juvenile muscular atrophy of the upper extremity. Eur J Radiol 13 : 215-219, 1991.*

Hirayama K, Toyokura Y, Tsubaki T : *Juvenile muscular atrophy of unilateral upper extremity : A new clinical entity. Seishin Shinkeigaku Zasshi 61 : 2190-2197, 1959.*

Hirayama K, Tsubaki T, Toyokura Y, Okinaka S : *Juvenile muscular atrophy of unilateral upper extremity. Neurology (Minneap) 13 : 373-380, 1963.*

Kitagawa M, Tashiro K, Kikuchi S, Matsuura T : *Correlation between clinical features and neuroradiological findings in juvenile muscular atrophy of unilateral upper extremity (Hirayama disease) with and without "tight dural canal in flexion". Rinsho Shinkeigaku 32 : 479-482, 1992.*

Meadows JC and Marsden CD : *A distal form of chronic spinal muscular atrophy. Neurology 19 : 53-58, 1969.*

Metcalf JC, Wood JB, Bertorini TE : *Benign focal amyotrophy : Metrizamide CT evidence of cord atrophy. Case report. Muscle Nerve 10 : 338-345, 1987.*

Mukai E, Sobue I, Muto T, Takahashi A, Soto S : *Abnormal radiological findings in juvenile-type distal and segmental muscular atrophy of upper extremities. Clin Neurol 25 : 620-626, 1985.*

Mulder DW, Rosenbaum RA, Layton DD : *Late progression of poliomyelitis or forme fruste amyotrophic lateral sclerosis ? Mayo Clin Proc 47 : 756-761, 1972.*

Mumenthaler M : *Myopathy in neuropathy. Muscle diseases. Proceedings of The International Symposium. Milan. pp 585, 1969.*

Murata K, Iwasaki S, Mano Y, Nakagawa H, Takayanagi T : *Pathogenesis of juvenile muscular atrophy of unilateral upper extremity in reference to venous con-*

*gestion of epidural space. Rinsho Shinkeigaku 32 : 213-216, 1992.*

O'Sullivan DJ and McLeod JG : *Distal chronic spinal muscular atrophy involving the hands. J Neurol Neurosurg Psychiatr 41 : 653-658, 1978.*

Prabhakar S, Chopra JS, Banerjee AK, Rana PVS : *Wasted leg syndrome : A clinical electrophysiological and histopathological study. Clin Neurol Neurosurg 83 : 19-28, 1981.*

Riggs JE, Schochet SS, Gutmann L : *Benign focal amyotrophy : Variant of chronic spinal muscular atrophy. Arch Neurol 41 : 678-679, 1984.*

Singh N, Sachdev KK, Susheela AK : *Juvenile muscular atrophy localized to arms. Arch Neurol 37 : 297-299, 1980.*

Spillane JD : *Localized neuritis of the shoulder girdle. Lancet 11 : 532, 1943.*

Spiro AJ : *Minipolymyoclonus : A neglected sign in childhood spinal muscular atrophy. Neurology 20 : 1124-1126, 1970.*

Tokumaru Y and Hirayama K : *Anterior shift of posterior lower cervical dura mater in patients with juvenile muscular atrophy of unilateral upper extremity. Rinsho Shinkeigaku 29 : 1237-1243, 1989.*