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Amyotrophic cervical myelopathy in adolescence

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Abstract

The clinical and radiological features in seven patients who had asymmetric muscular atrophy of the hand and forearm when young are reported and a new hypothesis for its aetiology is proposed. Investigation of body growth curves (a surrogate for velocity of arm growth) showed close relation between (*a*) the age when the body height increased most rapidly and the onset age of this disorder, and (*b*) the age when the rapid body growth period ended and the age when symptom progression ceased. Cervical radiological evidence is provided showing asymmetric anterior cord atrophy, disappearance of slackness of dorsal roots in neck extension, and anterior and lateral displacement of the lower cervical cord against the posterior aspects of the vertebral bodies during neck flexion. These results suggest that disproportionate shortening of the dorsal roots is further accentuated during the juvenile growth spurt, which determines the onset and self limited course of the condition, and that repeated neck flexion causes microtrauma and relative ischaemia of anterior horn cells, which finally results in atrophy of the muscles innervated by motoneurons with long axons. Predisposing anatomical factors are a straight neck due to lack of physiological cervical lordosis and the presence of foreshortened dorsal roots.

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Hirayama *et al*[1] and Takagi and Okabe,[2] were the first to report, independently in 1959, juvenile cases of unilateral muscular atrophy localised to the hand and forearm. Hirayama *et al* initially called this disorder juvenile muscular atrophy of unilateral upper extremity,[3] and later designated it juvenile non-progressive muscular atrophy localised in the hand and forearm.[4] Sobue *et al*[5] referred to it as distal localised muscular atrophy of upper extremities with juvenile onset, and thereafter rephrased it as segmental muscular atrophy of distal upper extremity with juvenile onset.[6]

The disorder is characterised by the following conditions: (*a*) muscular atrophy that insidiously appears in youth with a much higher incidence in males than in females; (*b*) distribution of muscular atrophy that is distinctly asymmetric and localised to the hand and forearm; (*c*) progression of muscular atrophy during the early onset period that becomes self limiting; and (*d*) most cases are sporadic and the patient's history is generally non-contributory. Most of the cases were Japanese[7–9] and only a few patients have been reported outside Japan.[10–19]

Recent advances in examination techniques have produced several characteristic findings for this disorder. Hashimoto *et al*[8] pointed out that a socalled straight neck due to lack of normal lordosis of the cervical spine was seen in plain radiographs. Matsumura *et al*[20] confirmed localised atrophy of the lower cervical cord in CT myelography. Yada *et al*[21] and Mukai *et al*[22] showed with dynamic myelography that when the neck was flexed, the dural sac was shifted anteriorly and the spinal cord was compressed and flattened between the posterior wall of the dural sac and the cervical vertebral body, suggesting that during neck flexion the dural sac and cord were overstretched. Since then, this "overstretching" has been thought to be one of the inductive factors of this disorder.

Although Hirayama *et al*[23] reported a necropsied case of this disorder, the true pathogenesis or causative mechanism is not known, and many questions concerning the clinical manifestations still remain unsolved.

In the present study, our aim was to clarify the causative mechanisms of this disorder by investigating seven patients in detail.

Patients and methods

Clinical neurological signs and symptoms, longitudinal growth curves (five patients only), and radiological examinations were analysed for seven patients (six males and one female) with asymmetric muscular atrophy localised to the hand and forearm. We constructed height increase curves to evaluate the relation between the onset of muscular atrophy and arm growth velocity, because arm growth is thought to parallel growth in body height. These longitudinal growth curves taken from our five patients were compared with the Japanese average curves obtained from data published in the 1989 Annual Report of the School Health Survey (SHS) by the Ministry of Education of Japan. Individual clinical neurological symptoms were superimposed on each curve.

Plain radiography, dynamic myelography,

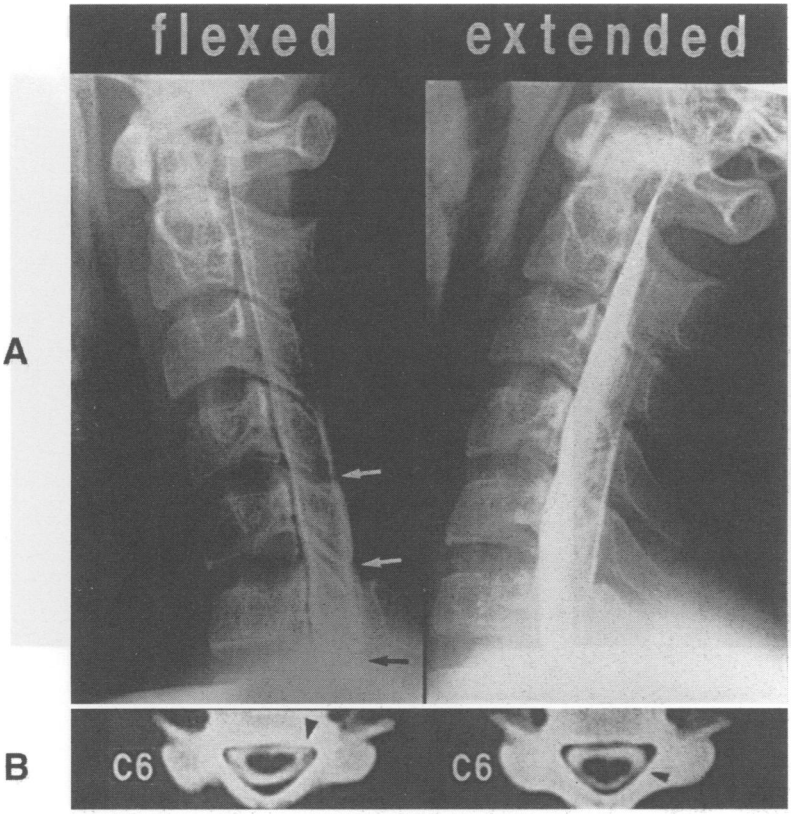

Figure 3 Myelograms and CT myelograms of case No 2 (the affected side is on the left). (A) Lateral view of myelograms. In neck flexion, the lower cervical cord and the posterior wall of the dural sac are shifted anteriorly, although the subdural space gets wider posterior to the cord (arrows). The posterior epidural space expands. In neck extension, the cord is moved posteriorly. As a result, the anterior subdural space gets wider and is filled more with contrast medium and the posterior epidural space is shrunk. (B) Transverse view of CT myelograms. In neck flexion, the cord is shifted anteriorly, being pulled to the affected side (arrow head) and flattened anteroposteriorly. As the posterior wall of the dural sac shifts anteriorly, the posterior epidural space expands. A space filled with the contrast medium between the cord and the posterior dura is seen. In neck extension, cuneiform atrophic cord is seen on the left side (arrow head) and the anterior subdural space is filled more with the contrast medium as the cord is moved posteriorly.

by pinioning in an accident. One year later, muscular atrophy also appeared in the right hand. He underwent cervical vertebral fusion at the age of 17 years and 9 months to limit the mobility of his neck. It was noted that he had played school volleyball and basketball between the ages of 13 and 15, during his peak growth period.

Figure 2B shows the longitudinal growth curve of a female patient (patient No 1). The average velocity curve for Japanese females (SHS, 1989) peaks at 11 years of age—that is, two years earlier than for males, and the annual average increase in height at the peak is about 8 cm, which is 2 cm less than for males. The slope of the average growth velocity curve is also less steep than that of males. Patient No 1 started to grow rapidly at the age of 11 to 12, although her body height was below average before the age of 11. At the age of 13, the patient was taller than average. Her growth velocity curve peaked at the age of 12 and the slope was steeper than that of the male average. She had played volleyball as school activity between the ages of 12 and 14. The initial symptom appeared as muscular atrophy at the age of 16—that is, four years after the peak of growth velocity. When her growth had almost ceased at the age of 18, the muscular atrophy ceased to worsen.

Figure 2C, D, and E show the growth curves of three other male patients (Nos 2, 4, and 7). The peak in their growth velocity curves occurred one to three years later than that of the normal average, latest in patient No 2. All five curves showed a relation between the rapid growth period and prolonged sports activities.

RADIOLOGICAL EXAMINATION

Plain radiography

Radiographs of the cervical spine showed no physiological lordosis in any of the seven patients, all of whom had socalled straight necks.

Dynamic myelography with the neck flexed and extended

Lateral view—When the neck was flexed, the lower part of the cervical cord and the posterior dura mater were moved forward and the posterior epidural space was enlarged (fig 3). There was some space filled with contrast medium between the spinal cord and the posterior dura mater. There was no evidence that the cord was being compressed by the dural sac.

Posteroanterior view—In normal subjects, when the neck was extended, there was slackness of the nerve roots; thus the C8 nerve root runs almost horizontally (fig 4C). The patients showed no slackness of the nerve roots during neck extension. The nerve roots were rather strained and ran downward, especially on the affected side. In addition, the paths of the nerve roots on the affected side were shorter than on the unaffected side, and the spinal cord deviated toward the shortened roots (fig 4A, B). When the neck was flexed forward, tension developed and stretched the lower cervical nerve roots. Furthermore, the nerve roots on the affected side were very much under strain because of the shortened paths; this resulted in the lower cervical cord being forcefully pulled to the affected side (fig 4A).

CT myelography

When the neck was extended, the lower part of the cervical cord was shifted slightly, laterally toward the affected side. The anterior subdural space was filled with contrast medium and a cuneiform atrophic cord was seen on the affected side (fig 3B, fig 5). When the neck was flexed, the lower part of the cervical cord was shifted anteriorly, anteroposteriorly flattened, and pulled sharply to the affected side (fig 3B). The posterior wall of the dural sac was also shifted. Therefore, the posterior epidural space was expanded. In patient No 2, there was a space filled with contrast medium between the spinal cord and the posterior dura mater. Therefore, no posterior compression of the spinal cord by the dura was found (fig 3B).

Patient No 6 had severe deformation of the dural sac (fig 6). In the neutral position, the spinal cord was flattened and deviated to the left side, especially at the level of the

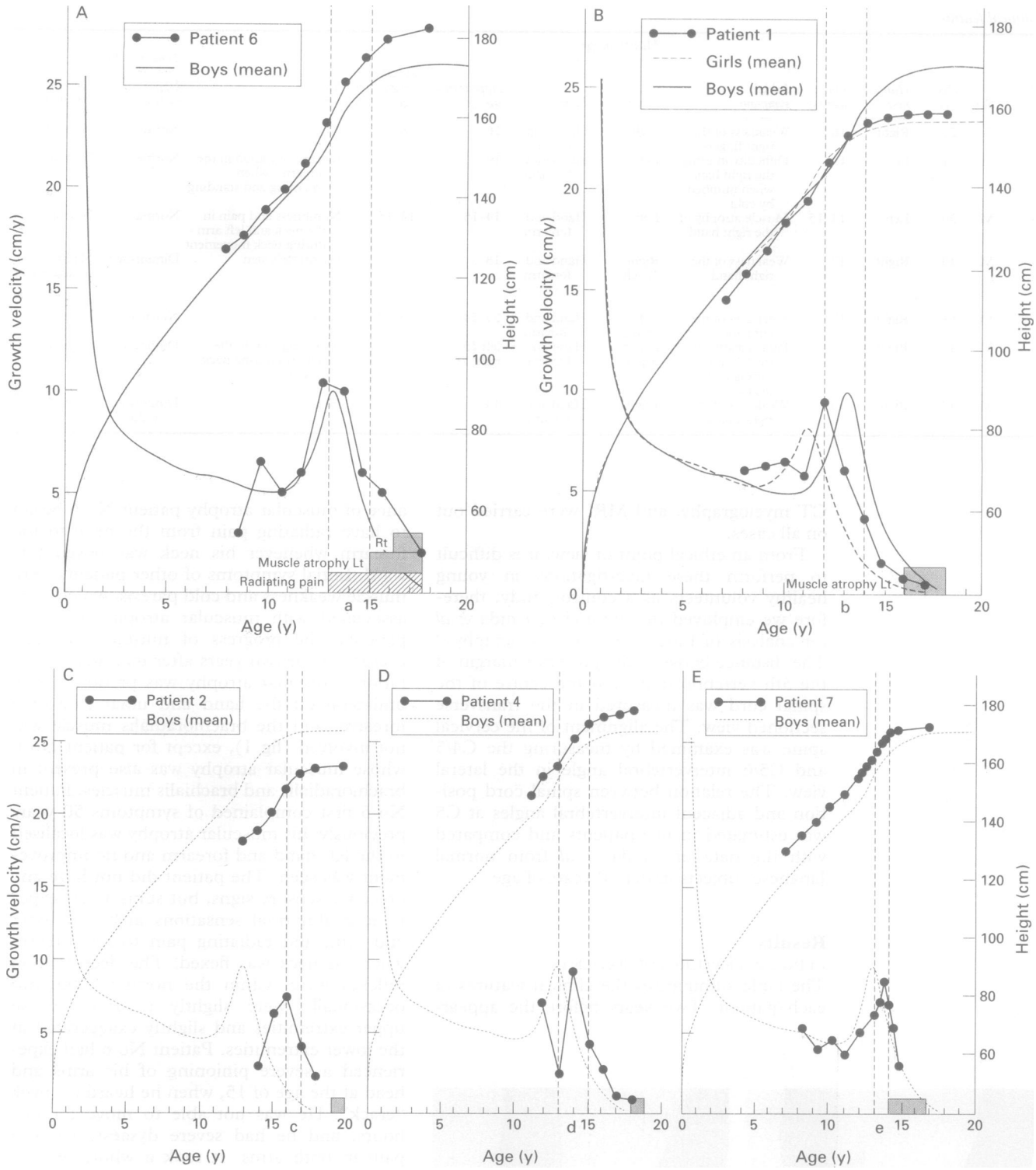

Figure 2 Individual growth velocity and longitudinal growth curves in relation to the appearance of each clinical symptom; thick line with filled circle, growth curve of each patient; thin line in A and B and the dashed line in C, D, and E, average growth curves for Japanese boys (from SHS, 1989); broken line in B, average growth curve for Japanese girls (from SHS, 1989); stippled bar indicates progression of muscular atrophy from onset to arrest; vertical dashed line indicates duration of strenuous sporting activity; a = basketball and volleyball; b = volleyball; c = boxing; d = long jump and sprint; e = sprint.

patient (patient No 6), who had remarkably rapid growth—that is, more than 10 cm growth per year—for two years between the ages of 12 and 14. His annual growth curve (growth velocity curve) at 12 years of age started to increase sharply. Compared with the average curve for Japanese males (from SHS, 1989), the peak period of the growth velocity curve was wider and the growth velocity after the age of 15 was also faster. When the growth velocity peaked at the age of 13, the initial symptom of radiating root pain in the left arm appeared. Muscular atrophy in the left hand was recognised two years after the peak in the velocity curve. At the age of 15 this patient's neck was forcefully flexed

Clinical features

Patient No	Sex	Age (y)	Handedness	Onset age (y)	Initial symptom	Muscle atrophy: Laterality	Muscle atrophy: Portion	Muscle atrophy: Appearance age (y)	Muscle atrophy: Arrest of progression age (y)	Sensory sign	Tendon reflex: Upper extremity	Tendon reflex: Lower extremity
1	F	20	Right	16	Weakness of the right fingers	Right	Hand and forearm	16	18	None	Normal	Normal
2	M	21	Left	19	Difficulty in using the right hand when numbed by cold	Left	Hand and forearm	19	20	Electric sensation in the forearms when squatting and standing	Normal	Normal
3	M	30	Left	14–15	Muscle atrophy of the right hand	Left	Hand and forearm	14–15	14–15	Numbness and pain in the neck and left arm during neck movement	Normal	Normal
4	M	19	Right	17	Weakness of the right hand	Right ⩾ left	Hand and forearm	18		Lhermitte's sign	Diminished	Slightly exaggerated
5	M	68	Right	17	Weakness of the left hand	Left ⩾ right	Hand and forearm	23–24	23–24	None	Normal	Normal
6	M	17	Right	13	Pain radiating in the left arm during neck flexion	Left > Right	Hand and forearm	left 15, right 16		Radiating pain in the left arm during neck flexion	Diminished	Normal
7	M	17	Right	13	Weakness of the right thumb	Right	Hand and forearm	14		None	Diminished (right)	Normal

CT myelography, and MRI were carried out on all cases.

From an ethical point of view, it is difficult to perform these investigations in young healthy volunteers as a control study; therefore we employed the method of Ishida *et al* for analysis of functional CT myelography.[24] The distance between the posterior margin of the 5th vertebral body and the centre of the spinal cord was measured in the transverse sectioned view. The alignment of the cervical spine was examined by measuring the C4/5 and C5/6 intervertebral angle in the lateral view. The relation between spinal cord position and adjacent intervertebral angles at C5 was estimated in five patients and compared with the data of Ishida *et al* from normal Japanese subjects under 30 years of age.

Results

CLINICAL FEATURES OF PATIENTS

The table summarises the clinical features of each patient. Two years before the appearance of muscular atrophy patient No 6 began to have radiating pain from the neck to the forearm whenever his neck was flexed forward. Initial symptoms of other patients were muscle weakness and cold paresis, which were associated with muscular atrophy. In these patients, the progress of muscular atrophy ceased one or two years after its initial recognition. Muscular atrophy was predominantly unilateral on the hand and ulnar side of a forearm, and the brachioradialis muscle was not involved (fig 1), except for patient No 6 whose muscular atrophy was also present in brachioradialis and brachialis muscles. Patient No 5 first complained of symptoms 50 years previously: his muscular atrophy was localised to the left hand and forearm and no improvement was seen. The patient did not have any objective sensory signs, but subjectively experienced abnormal sensations in his shoulder and arm, and radiating pain to his forearm when his neck was flexed. The deep tendon reflexes were within the normal range but occasionally were slightly reduced in the upper extremities and slightly exaggerated in the lower extremities. Patient No 6 had experienced a severe pinioning of his arms and head at the age of 15, when he heard his neck "crack". He was not able to move for two hours, and he had severe dysaesthesia and pain in both arms. It took a whole day for both arms to regain normal function. The remaining six patients presented here had no history of trauma. It was noted that six of these seven patients had participated in strenuous sporting activities at school in their teens—that is, during their rapid growth period—and they thereafter developed muscular atrophy within one to three years despite discontinuing their sports (fig 2).

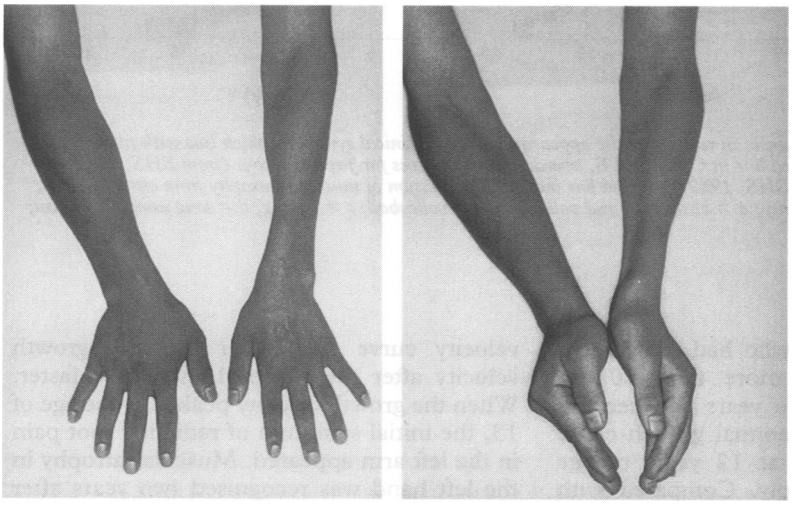

Figure 1 Muscular atrophy localised to the hand and forearm, predominantly on the left (case 2).

RELATION BETWEEN ARM GROWTH AND ONSET OF MUSCULAR ATROPHY

Rapid growth in height usually accompanies rapid arm and leg growth. Theoretically arm span is nearly equal to body height. Figure 2A shows the longitudinal growth curve of a male

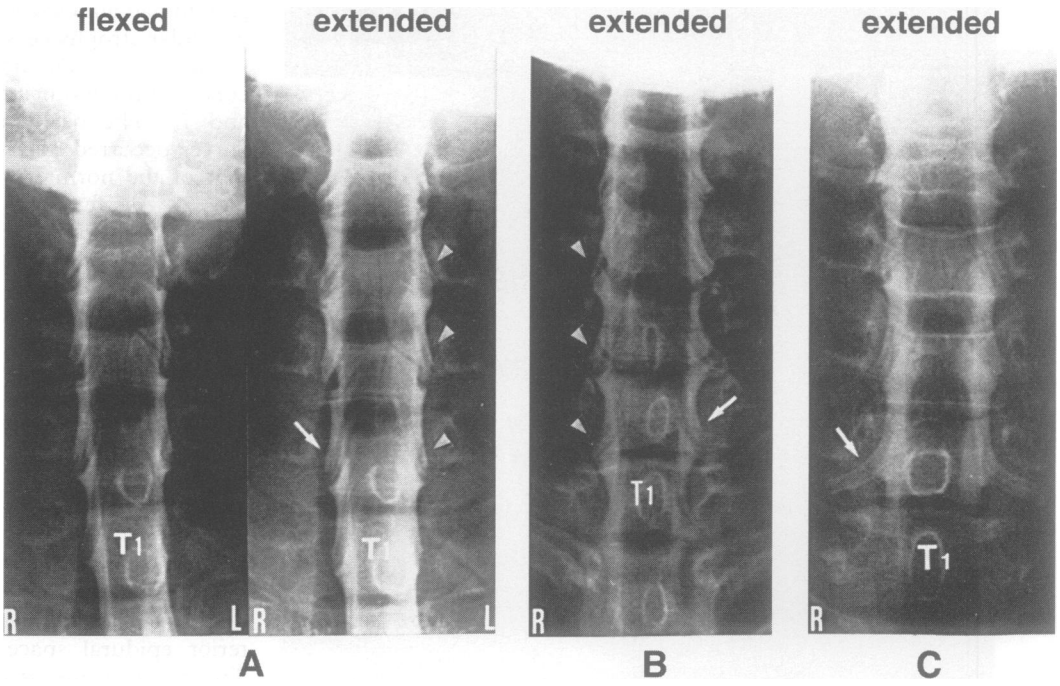

Figure 4 Posteroanterior view myelograms. (A) Case No 2 (affected side is on the left). In neck extension, the 6th, 7th, and 8th nerve roots on the left (three white arrow heads) are very strained, and the cord has a traction at these points. The 7th and 8th nerves (white arrow) on the right are also slightly strained. In neck flexion, the nerve roots on the left are extremely strained and the cord is moved more laterally to the left. (B) Case No 1 (the affected side is on the right). In neck extension, slackness of the nerve roots on the left is almost normal (the arrow on the left indicates the 8th nerve). On the other hand, the slackness is lost on the nerve roots on the right side, and the cord is pulled towards the right (three white arrow heads). (C) Normal subject aged 30. The nerve roots have normal slackness in neck extension and run almost horizontally, especially the 8th root (arrow).

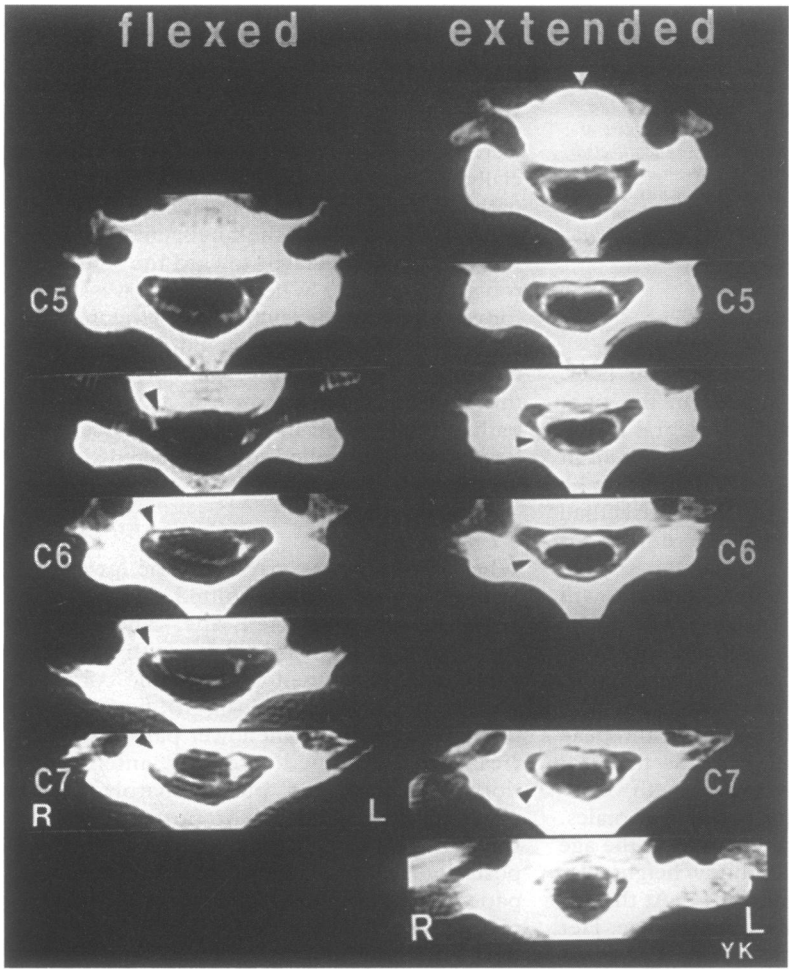

Figure 5 CT myelograms of case No 1 (the affected side is on the right). In neck extension, the lower cervical cord is shifted laterally to the right. Cuneiformed atrophic cord is seen on the affected side (arrow heads). The anterior subdural space is filled more with the contrast medium as the cord is moved posteriorly. In neck flexion, the cord is shifted anteriorly, sharply pulled to the affected side (arrow heads), and flattened anteroposteriorly. As the posterior wall of the dural sac shifts anteriorly, the posterior epidural space expands.

C5–7 vertebral bodies (arrow head fig 6) and there was a space filled with contrast medium anterior to the cord. When the neck was flexed, the cord was shifted anteriorly, the right side of the spinal cord was rotated in the right anterior direction around the left entry zone of the C5–6 dorsal roots, and the dural sac was constricted along the posterior midline portion. No subdural space was seen on the right side anterior to the cord, especially at the C5–7 vertebral bodies, and the right posterior subdural space was filled with contrast medium. Therefore, it was confirmed that there was no compression of the spinal cord by the posterior dura mater. Figure 7 explains how the cord was rotated and the dural sac deformed.

Figure 8 shows the relation between spinal cord position and adjacent intervertebral angles at C5 in five patients. The slopes of the regression lines for those patients under 30 years old were steeper than those for normal controls obtained from the study of Ishida *et al.*[24] These results indicate that the degree of anterior shift of the spinal cord in neck flexion was more prominent in the patients than in normal young subjects.

MRI

Asymmetric cord atrophy with dominance on the same side as the muscular atrophy was found at the C5–C7 level by MRI. When MRI was taken with the neck ventroflexed, an anterior shift of the cord and dural sac was seen and a flattened cord contacted the ventral border of the vertebral body. In addition, gadolinium-GTPA enhanced MRI in T1 images often showed a highly enhanced venous plexus in the posterior epidural space from the lower cervical to the thoratic vertebral body during neck flexion.

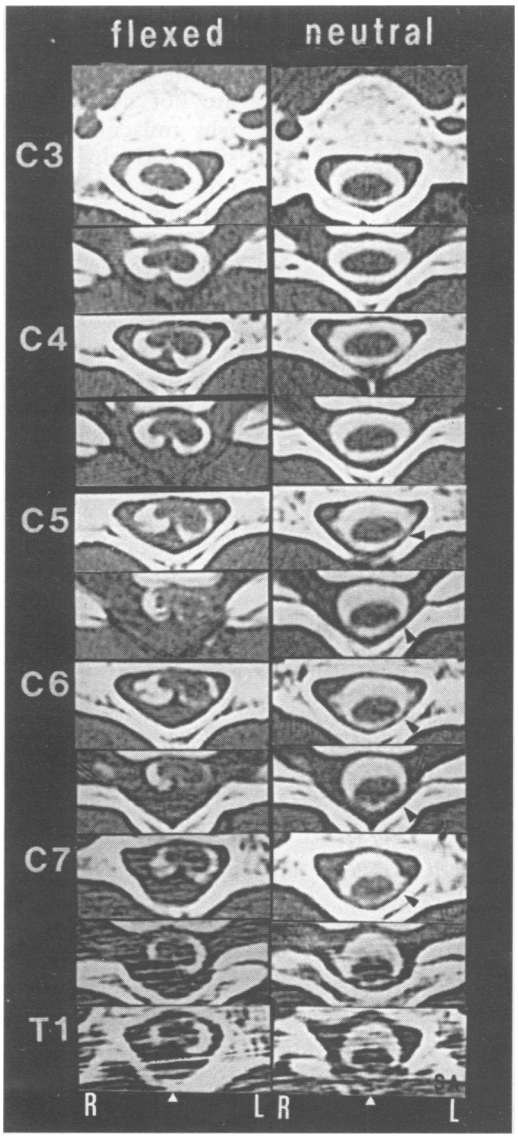

Figure 6 CT myelograms of case No 6. In neck flexion, the cord is shifted anteriorly and the right side of the spinal cord is rotated in the right anterior direction. There is no subdural space anterior to the cord because it touches the posterior border of the vertebral body at the level of C 5–7. The right posterior subdural space is filled with the contrast medium. The dural sac is constricted along the midline of the posterior wall. In the neutral position, the cord is situated posteriorly and deviates to the left (arrow heads) and a space filled more with contrast medium is seen anterior to the cord.

Discussion

In dynamic myelography of patients with amyotrophic cervical myelopathy, anterior shifting of the posterior dura mater, together with flattening of the lower cervical cord between the posterior wall of the dural sac and the vertebral body, were found when the neck was flexed.[21 22] Thus some investigators have referred to this condition as flexion myelopathy,[25] overstretch mechanism,[21] or tight dural canal in flexion[25] and considered it to be a part of the pathogenesis of flexion myelopathy. Both the spinal cord and dural sac are overstretched in a vertical direction when the neck is flexed, and then the spinal cord is pushed against the posterior vertebral body. The cause of this disorder is thus explained by the imbalance between development of the cervical cord and the spine. This, in turn, causes strain on the dural sac during neck flexion, which results in the spinal cord being pushed anteriorly. In most patients with this disorder, however, there was an actual space between the anteriorly displaced cord and the posterior dura mater, and compression of the cord from the posterior dura was rarely seen.

Figure 9 schematises the dynamic changes in the cervical cord and dural sac on flexion and extension of the neck. During neck extension in normal subjects, the spinal cord was shifted caudally, and the dorsal roots and the blood vessels were slack (fig 9A). When the neck was flexed forward in normal subjects, the dorsal roots were extended in an anterorostral direction with an anterorostral shift of the cervical cord (fig 9A).[26] In fact, the cervical spinal cord shifted anteriorly in flexion and posteriorly in extension of the neck, and was flattened at the midcervical level in flexion in normal young subjects.[24] In the present study, however, we found that the patients' dorsal roots did not show slackness when the neck was extended (fig 9B). When the patient's neck was flexed, the dorsal roots were insufficiently extended due to lack of slackness in extension. This resulted in the dorsal roots becoming short in relation to the spinal cord elongation during neck flexion (fig 9B).

The spinal cord connects with the dural sac by means of the denticulate ligaments and the septum subarachnoidale posterius, and floats in the CSF. In these patients, the relatively short dorsal roots pull the spinal cord anteriorly beyond the normal degree of shift when the spinal cord is stretched during neck flexion, even over a small range. Because the spinal cord is tightly connected to the dural sac on the posterior side by the septum subarachnoidale posterius, this shift of the spinal cord also makes the posterior wall of the dural sac move forward. Because these patients have asymmetric shortening on the right and left dorsal roots, the cord is then pulled to the side where the shortening is most severe (fig 9B). Finally, a large and extremely asymmetric difference of forward and upward force would generate a pronounced asymmetric forward shift and upward stretch of the cord, which could result in a serious situation as seen in patient No 6—that is, rotation of the cord and deformation of the dural sac (figs 6 and 7).

Dilatation of the venous plexus appears in these patients during neck flexion, because the pressure inside the posterior epidural space decreases when the posterior wall of the dural sac moves forward (fig 9B).

We think that the dorsal roots become short because the growth of the cervical roots does not keep up with the rate of skeletal arm growth during the rapid growth period in some young people. Reid[27] confirmed, in cadavers, that lateral movements may be seen in the cervical roots on abduction of the arm at the shoulder or on pulling the arm downward. Therefore, this relative shortness of the C5-Th1 dorsal roots during the rapid growth period pulls the lower cervical cord anteriorly when the neck is flexed. As a result, the spinal cord is compressed by the vertebral body, and chronic degenerative changes can occur in the anterior horns.

This disorder occurs mainly in young males who are growing rapidly. In such people, not only body height, but also arm and leg lengths increase considerably. In addition, it has been

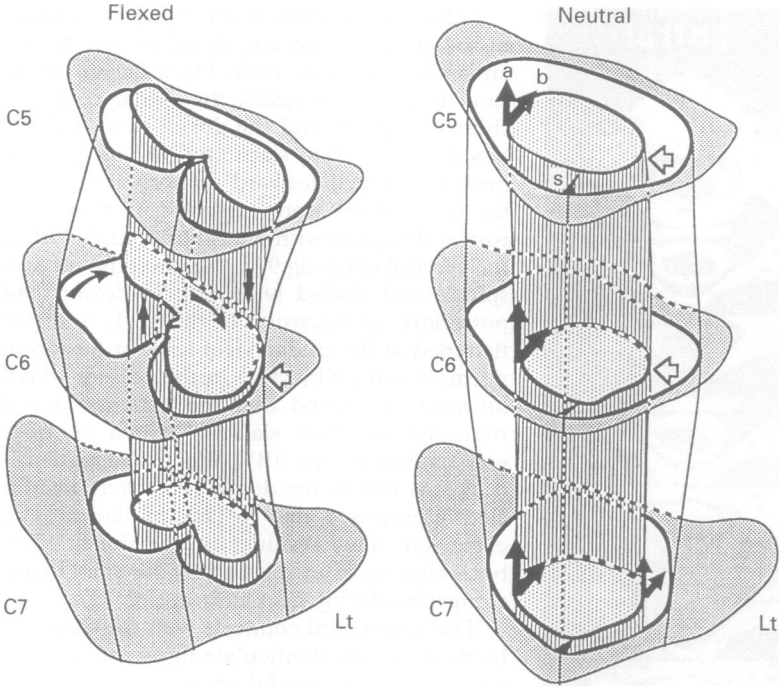

Figure 7 Schematic diagram of dynamic change of the cord and the dural sac during neck flexion in case No 6. When the neck is flexed forward from the neutral position, upward extension force (a) and forward traction force (b) are generated over the right and left dorsal root entry zone of the cervical cord according to their root extensibilities. In patient No 6, the extensibility of the roots is asymmetric—that is, left roots are not able to extend any more because they are too short (indicated by open arrows). During neck flexion, the left dorsal roots at the C5th and 6th vertebral bodies do not move. The right side of the cord is able to extend upward and at the same time to rotate in the right anterior direction (indicated by arrows) around the immovable left nerve root entry zone (open arrow). The dural sac is, consequently, constricted along the posterior midline because the spinal cord is tightly connected with the dural sac on the posterior side by the septum subarachnoidale posterius (s).

reported that in this growing period, the ratio of sitting height to height is the smallest. From this point of view, the present results are not consistent with the hypothesis of the overstretching of the spinal cord due to imbalanced development between the cord and spine during the growing period.[21]

The comparative analysis between the growth curve and age of onset showed that this disorder occurs at an age when the length of the arms, as well as body height, develop most rapidly. Muscular atrophy generally appeared two to four years after the onset of the initial symptoms, and stopped progressing when rapid growth was over. The data from SHS showed that young Japanese men grow most rapidly at age 13, and rapid growth continues until 18–19 years of age. This period is considered to be the latent period of recognisable muscular atrophy. We found 49 papers written in Japan on this disorder. Among them, the age at onset was recorded for 160 male patients, 80% of whom had initial symptoms before they were 19 years old. As in most cases the initial symptom is muscular atrophy of the hand, clinical features of the disorder are established when muscular atrophy is noticed.

The growth peak of women is two years earlier than in men. The largest increase in annual body height is about 8 cm, 2 cm less for women than for men, and the slope of the growth velocity curve of women is not as steep as for men. Therefore, women are rarely afflicted with this type of muscular atrophy.

The growth rate of the left and right arm are not usually uniform. This may explain why muscular atrophy can appear on either side, and why the major symptoms appear on only one side. According to one report,[4] more patients have muscular atrophy in the right arm than in the left, which might be related to faster growth of the right arm. No relations have been found to the patients' handedness, however.[8 19]

There are several reports on families with histories of this disorder,[7 9 23] not surprising as growth patterns are largely dependent on hereditary factors.

Because of its benign course, studies on the pathology of this disorder have rarely been performed. Hirayama *et al*[23] first reported pathological findings on a patient with this disease who died of lung cancer. Lesions existed only in the anterior horns of the bilateral lower cervical cord, and showed shrinkage and necrosis, various degrees of degeneration of nerve cells, and mild gliosis. They suggested that the lesions were produced by a consequent circulatory insufficiency of the cervical cord. Pushing the

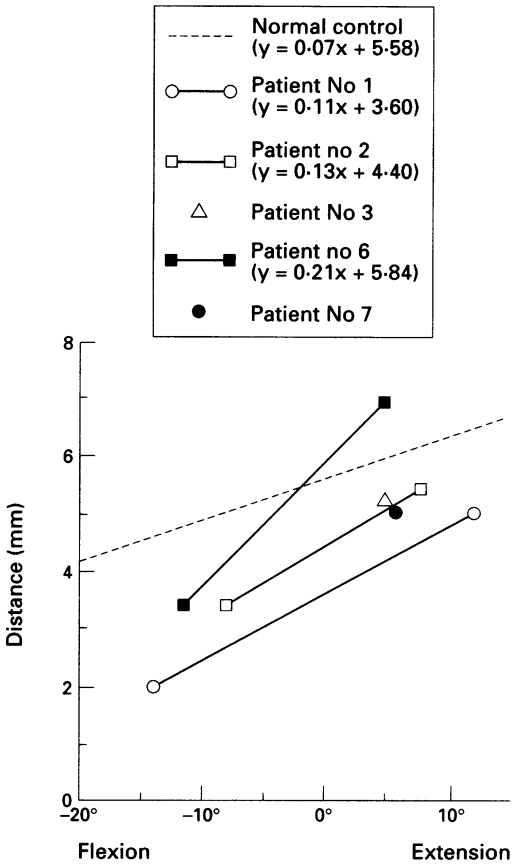

Figure 8 Relation between adjacent intervertebral angles and location of the spinal cord at the level of C5 in patients under 30 years old. X = Sum of angles at C4/5 and C5/6; Y = values of distance to the vertebral body at C5. The slopes of the regression lines in the three patients are larger than those of normal controls from the data of Ishida et al.[24] The lower cervical cords in the patients are situated more anterior than in normal controls, especially during neck flexion.

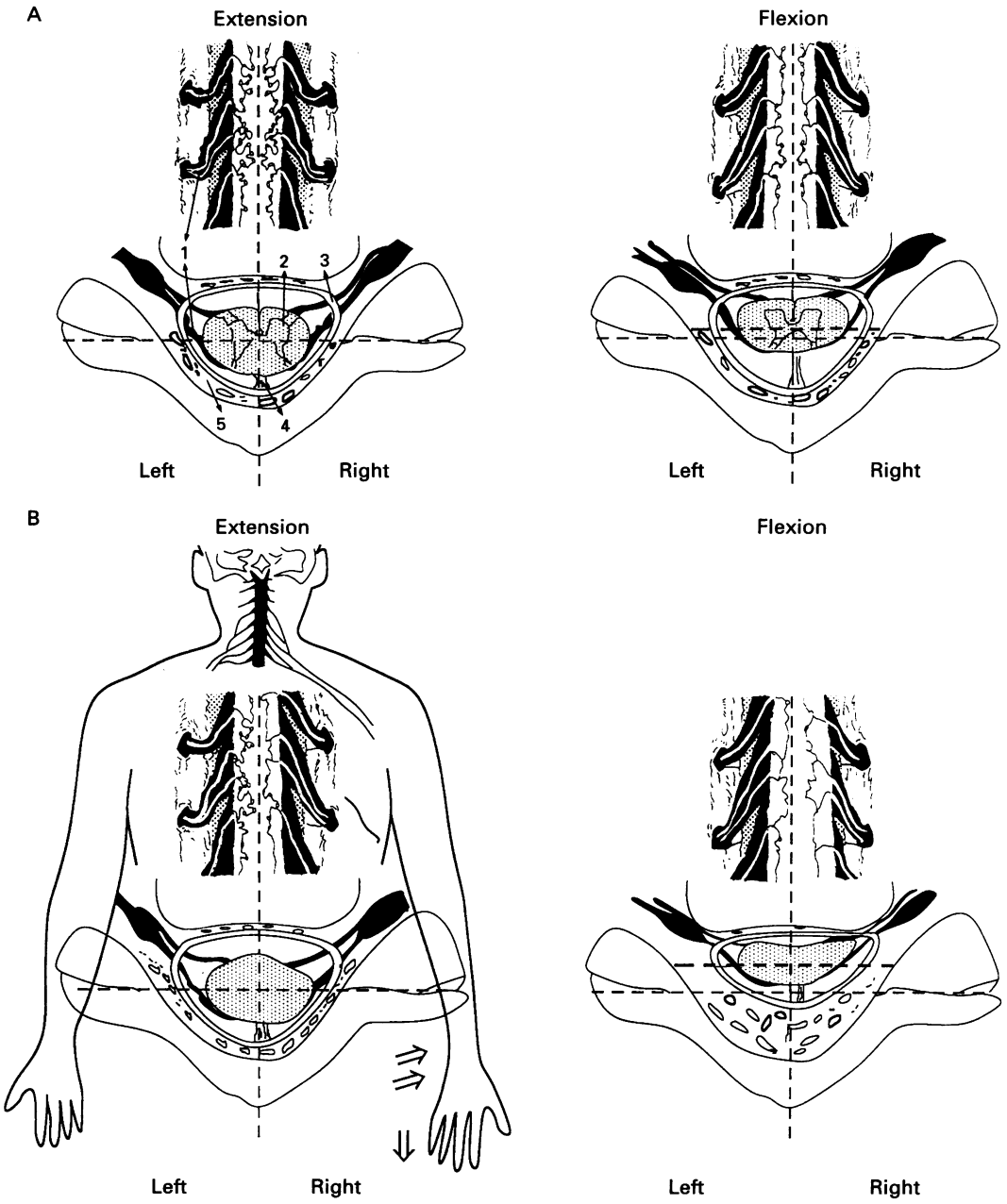

Figure 9 Schematic drawings of the mechanism of dynamic change of the cervical cord and dural sac related to dorsal roots during neck extension and flexion. Vertical broken line shows the midline of the body. The longer horizontal broken line shows the centre of the cord during neck extension. The shorter horizontal broken line indicates the cord shifted anteriorly during flexion. (A) Normal subject; during neck extension the dorsal roots, cord, and dura in the cervical canal are slack. On neck flexion, the roots, cord, and dura are drawn out; elongation of the dorsal roots and cord is permitted by the slack present in neck extension (1 = dorsal root; 2 = spinal cord; 3 = dural sac; 4 = septum subarachnoidale; 5 = epidural space). (B) Patient affected on the right side (arrows); during neck extension, slackness of the dorsal roots disappears on the affected side (right) in which the arm grows faster than on the other side. On neck flexion, the right dorsal roots cannot extend, and this causes the spinal cord to be drawn anteriorly to the right.

anterior aspect of the spinal cord to the vertebral body might result in chronic compressive mild circulatory changes to the anterior horn tissues, which are the most vulnerable to ischaemia.

In general, chronic functional damage to neurons for two or more years will cause dysfunction of muscles, especially when they are innervated by long axons; these axons have the greatest demand for resupply, and their distal regions are the most remote areas of somal irrigation.[28] This is presumably the reason why muscular atrophy is localised in the hand and the ulnar side of the forearm, as socalled oblique amyotrophy.[29] Electromyographic findings showed typical neurogenic changes not only in the atrophied muscles, but also in the non-atrophic homonymous muscles. Such abnormal changes were not found in the brachioradial, extensor calpi radialis longus, pronator teres, and pectoralis major muscles, despite their C5–7 segmental innervations.[30 31] This could be explained by the fact that the motor points of these muscles are located in a more proximal portion of the upper extremity.

Patients occasionally reported subjective sensory symptoms. Our patient No 6 had radicular pain as an initial symptom when the neck was flexed forward. This pain preceded a recognised muscular atrophy by about two years. There are some reports of radicular pain during neck flexion, and of Lhermitte's sign.[32] We think that the radicular stimulating sign is due to traction of lower cervical rootlets or roots when the neck is flexed. In this situation sensory disturbances have not yet been studied in detail, because the severity of organic changes of the dorsal rootlets is usually mild and objective signs are few.

In patient No 6, myelography and CT myelography were performed four and a half years after the initial symptoms, and depicted the spinal cord showing extreme rotation in a

right anterior direction around the shortened left and fixed C5–6 dorsal root entry zone, and the anterior right half of the cord touched the vertebral body (fig 6). These findings also indicated the presence of muscular atrophy in the right hand, which appeared later than the left hand atrophy. In this patient in the earliest stages, damage to the left anterior horns developed from the severe shift of the cord to the left. As growth continued, the left dorsal roots could not extend when the neck was flexed, and the cord began to rotate to the right around these left unextensible roots (fig 6). There is a possibility that the forceful pinioning, which occurred, when the patient was 15 years old, triggered rotation of the cord and then deformation of the dural sac. The degree of shift and rotation of the cord could depend on the balance between the arm growth process and the relative shortening of the roots.

The cause of this disorder depends on how the skeleton of the body grows during the adolescent period. We think that the incidence will be higher in persons whose growth is initially slow but which suddenly increases later as in our patient No 1. Persons with straight necks, who do not have physiological cervical lordosis, are more prone because their roots might be already stretched. The occurrence of this disorder might be prevented by avoiding physical exercises that cause frequent and violent flexion of the neck. Strenuous exercise in sports was often noted in patient case histories.[8 16 17] From our studies, we propose that participation in sports during the rapid growth period is one of several high risk factors for this disease. The risk of appearance of this disorder will decrease after skeletal growth slows down and dorsal root slackness develops sufficiently. In patient No 5, muscular atrophy has not improved for 50 years, but the patient is capable of performing simple isotonic finger movements. In principle, damaged neurons can be repaired if degenerative changes of the distal parts of axons are reversible. Therefore, for both high risk persons and people who have had asymmetric muscular atrophy of the hand and forearm, forward flexion of the neck should be avoided until skeletal growth has been completed.

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