Cervical Spondylotic Amyotrophy Treated by Anterior Decompression —Three Case Reports—

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Abstract

Cervical spondylotic amyotrophy (CSA) is a rare type of cervical spondylotic disorder. The surgical treatment of CSA is controversial. We treated three patients with the proximal type of CSA by anterior decompression. Three men (65, 61, and 58 years old) presented with CSA manifesting as unilateral muscle weakness and atrophy in the deltoid and biceps muscles without significant sensory deficit. Preoperative neuroradiological examinations revealed anterolateral spinal cord compression in one patient and ventral root compression in two patients at the C4-5 and C5-6 spaces. Magnetic resonance (MR) imaging showed no abnormal intramedullary signal intensity in any patient. Vertebroto my deviated to the lesion side was performed to provide a better view of the laterally situated osteophytes. Anterior decompression was focused on the paramedial to lateral area, and further foraminotomy was performed according to the radiological findings. After decompression, intervertebral instrumentation was performed using titanium cervical cages. Two of the patients had good recovery of motor strength and one had moderate recovery despite persistent motor atrophy. Surgical intervention is effective in patients with CSA despite severe muscle atrophy unless MR imaging shows the presence of severe degenerative intramedullary lesion.

Key words: cervical spondylotic amyotrophy, anterior decompression, intervertebral cage

Introduction

Cervical spondylotic amyotrophy (CSA) or dissociated motor loss in the upper extremities is a rare form of cervical spondylosis which is characterized by severe muscle atrophy and weakness in the upper extremities without significant sensory deficits or myelopathy.^{1,9,10,17)} The clinical characteristics, electrophysiological findings, and neuroradiological findings of CSA have been well documented.^{7,12,13)} CSA can be divided into two types according to the affected muscles in the upper limb.¹³⁾ The proximal type (affected levels at C-5 and C-6) manifests as muscle atrophy and weakness predominantly in the unilateral scapular, deltoid, and biceps muscles, and the distal type (affected levels at C-7, C-8, and T-1) in the flexor carpi ulnaris, abductor pollicis brevis, and abductor digiti minimi muscles. Both types occur predominantly in males in middle to old age. Deep tendon reflexes in the upper extremity are diminished in both types. CSA affects mostly the unilateral

upper extremity but electromyography (EMG) shows neurogenic changes with fasciculation in both the affected and contralateral muscles. However, surgical treatment of CSA has only been described in a few cases because differentiation from motor neuron diseases seems to be difficult.^{8,10,12)} The prognostic clinical features and radiological findings, and the indications for surgery and the treatment modality have not been fully addressed.

We recently treated three patients with the proximal type of CSA by anterior decompression and fusion with titanium cervical cages, resulting in good and moderate outcomes.

Case Reports

Case 1: A 65-year-old male with a medical history of aortic valve disease experienced transient pain in the right shoulder which gradually developed into the inability to abduct and elevate the right arm in June 2003. At the same time, he noticed muscle atrophy in the right shoulder girdle. He was examined by a neurologist of our institution and admitted

to our neurosurgical department because of the presence of cervical spondylosis on August 29, 2003.

Neurological examination showed motor weakness of the right deltoid (manual muscle test [MMT]: 3), biceps (MMT: 3), triceps (MMT: 4), and brachioradialis (MMT: 4) muscles. These muscles and the right supraspinatus and infraspinatus muscles were severely atrophic. Deep tendon reflexes in the right biceps, brachioradialis, and triceps muscles were diminished, but other reflexes were normal. He did not complain of any sensory disturbance on admission, but careful neurological examination revealed subtle diminishment of touch and pain sensations in the C-5 and C-6 dermatomes of the right arm. EMG showed bilateral C-5 and C-6 segmental neurogenic changes with fibrillation, with predominance on the right.

Cervical radiography showed bilateral posterolateral spur formations at the C4-5 and C5-6 spaces. Sagittal T₂-weighted magnetic resonance (MR) imaging showed cord compression at the C4-5 and C5-6 spaces without intramedullary hyperintensity (Fig. 1A). Axial T₂-weighted MR imaging and computed tomography (CT) myelography revealed compression of the right side of the anterolateral spinal cord at the C4-5 space and the left side at the C5-6 space (Fig. 1B, C). The diagnosis was CSA at the C4-5 and C5-6 spaces.

Surgery was performed on September 11, 2003. The removal of the posterolateral osteophyte of the right C4-5 space was more important, so the neck skin incision was performed along the left sternocleidomastoid muscle. Vertebroto my at the C4-5 space was intentionally deviated 5 mm to the right of the midline and vertebroto my at the C5-6 space was performed to access the posterolateral osteophytes on each side. The compressing spur was removed under the operating microscope until the beginning of the intervertebral foramen was widened. After decompression, the C-4 and C-5, and C-5 and C-6 vertebrae were fused with titanium cages (M-cage; Blackstone Medical Inc., Springfield, Mass., U.S.A.). Postoperative neuroradiological examinations confirmed the relief of cord compression at both levels (Fig. 1D, E). After the surgery, the patient showed rapid improvement of motor weakness, and at 3 months after the surgery he had fairly recovered right arm abduction and elevation despite persistent muscle atrophy (deltoid: 4+, biceps: 4+, triceps: 4+).

Case 2: A 61-year-old male without any contributory medical history began to feel clumsiness in the left hand in March 2004. At the same time, he began to feel difficulty in lifting the left hand. He was examined by a neurologist in our institution on April 3,

Fig. 1 Case 1. A: Sagittal T₂-weighted magnetic resonance (MR) image showing multisegmental cord compression at the C4-5 and C5-6 spaces. B: Axial T₂-weighted MR images at the C4-5 (upper) and C5-6 spaces (lower) showing no abnormal signal intensity in the intramedullary spinal cord. C: Computed tomography (CT) myelograms showing flattening of the right anterolateral spinal cord at the C4-5 space (upper) and left anterolateral spinal cord at the C5-6 space (lower). D: Postoperative sagittal T₂-weighted MR image showing no segmental spondylotic changes. E: Postoperative three-dimensional bone CT (upper) and axial bone CT scans (lower) at the C4-5 space showing the titanium cervical cage and removal of the right posterolateral osteophyte at the C4-5 space.

2004 and referred to the neurosurgical department. He was admitted on May 12, 2004.

Neurological examination showed muscle weakness in the left deltoid (MMT: 2) and biceps (MMT: 3) muscles with severe atrophy. Deep tendon reflexes in the left biceps and triceps muscles were diminished without pathological reflexes. He sometimes felt abnormal sensations in his left thumb but sensory examinations did not detect any objective abnormalities. EMG showed neurogenic patterns in the left deltoid and biceps muscles. Cervical radiography showed bilateral hypertrophy of the unco-vertebral joints of the C-5 and C-6 vertebrae. T₂-weighted MR imaging showed mild multisegmental spinal cord compression at the C3-4, C4-5, and

Fig. 2 Case 2. A: Sagittal T₂-weighted magnetic resonance (MR) image showing multisegmental cervical spondylotic changes from the C3-4 to C5-6 spaces. B, C: Computed tomography (CT) myelograms showing axillary pouch deficits at the left C4-5 (arrow) and C5-6 (arrowhead) spaces (B) with narrowing of the intervertebral foramen at the left C4-5 (arrow) (C). D: Postoperative sagittal T₂-weighted MR image showing no segmental spondylotic changes. E: Postoperative three-dimensional bone CT (upper) and axial bone CT scans (lower) at the C4-5 space showing the titanium cervical cage and removal of the right posterolateral osteophyte and opening of the left intervertebral foramen at the C4-5 space.

C5-6 spaces without intramedullary hyperintensity (Fig. 2A). CT myelography disclosed axillary pouch deficits in the left C4-5 and C5-6 spaces (Fig. 2B) with narrowing of the intervertebral foramen at C4-5 (Fig. 2C).

Surgery was performed on May 27, 2004. After right side neck skin incision, vertebrotomies were performed 5 mm offset to the left at the C4-5 and C5-6 spaces. After removing the left posterolateral osteophytes, anterior foraminotomies were performed at the left C-5 and C-6 roots. Anterior fusion using titanium cages (AFFINITYTM; Medtronic Sofamor Danek, Inc., Memphis, Tenn., U.S.A.) was then performed of the C-4 and C-5, and C-5 and C-6 vertebrae. Postoperative radiography showed improvement of cord compression and widened intervertebral foramina (Fig. 2D, E). The motor weakness

in the left arm gradually improved and fairly recovered at 3 months after the surgery despite persistent muscle atrophy (deltoid: 4+, biceps: 4+).

Case 3: A 58-year-old male without any contributory medical history gradually lost the ability to abduct and lift his left arm in March 2003. He was treated by a private clinician under a diagnosis of periarthritis of the shoulder. However, his motor weakness worsened and he could not wash his face by hand. Examination at our neurosurgical department established a diagnosis of cervical spondylosis. He was admitted on August 19, 2004.

Neurological examination showed motor weakness of the left deltoid (MMT: 2), biceps (MMT: 2), and brachioradialis (MMT: 3) muscles with severe atrophy. The left supraspinatus and infraspinatus muscles also showed atrophy. Deep tendon reflexes in the left biceps, triceps, and brachioradialis muscles were diminished without pathological reflexes. Sensory examination showed no abnormalities. EMG showed bilateral C-5 and C-6 segmental neurogenic patterns with fibrillation, with left side predominance. Cervical radiography showed bilateral hypertrophy of the unco-vertebral joints at the C-4, C-5, and C-6 vertebrae. T₂-weighted MR imaging showed mild multisegmental cord compression from the C3-4 to C5-6 spaces (Fig. 3A, B). CT myelography disclosed left ventral root compression at the C4-5 and C5-6 spaces (Fig. 3C).

Surgery was performed on September 2, 2004. After right side neck skin incision, vertebrotomies were performed 5 mm offset to the left at the C4-5 and C5-6 spaces under the operating microscope. Foraminotomies were performed at the C-5 and C-6 roots. Anterior fusion using titanium cages (AFFINITYTM) was performed at the C-4 and C-5, and C-5 and C-6 vertebrae. Postoperative neuroradiological studies confirmed widened foramina at the C4-5 and C5-6 spaces (Fig. 3D, E). The patient showed gradual improvement and could wash his face because of good recovery of the muscle power in the left brachioradialis muscle, and he could lift his left arm to 45 degrees from the horizontal despite persistent muscle atrophy at 3 months after treatment.

Discussion

Whether CSA is associated with selective damage to the ventral roots or the anterior horn of the spinal cord remains unclear.^{7,17)} The first autopsy case of “dissociated motor loss in the upper extremity with cervical spondylosis” was attributed to selective intradural compression of the ventral roots due to posterolateral osteophytes.⁹⁾ CSA has also been con-

Fig. 3 Case 3. A: Sagittal T₂-weighted magnetic resonance (MR) image showing multisegmental spondylotic changes from the C3-4 to C5-6 spaces. B: Axial T₂-weighted MR images at the C4-5 (upper) and C5-6 spaces (lower) showing no abnormal signal intensity in the intramedullary spinal cord. C: Computed tomography (CT) myelograms showing left nerve root compression at the C4-5 (upper) and C5-6 spaces (lower). D: Postoperative sagittal T₂-weighted MR image showing no segmental spondylotic spurs. E: Postoperative three-dimensional bone CT (upper) and axial bone CT scans (lower) at the C4-5 space showing the titanium cervical cage and removal of the right posterolateral bony spur at the C4-5 space.

sidered to result from selective damage of the anterior horns due to circulatory disturbance in the territories of the spinal arteries,¹⁷⁾ and both the ventral roots and anterior horns were involved by paramedian compression.⁴⁾ Two of the present cases of CSA were caused by ventral root compression and the other case by spinal cord compression. Therefore, we assume that the pathogenesis of CSA can originate at either of these sites.

The clinical features of CSA such as muscle weakness and atrophy, and even fasciculation of the upper extremities in the absence of sensory symptoms, resemble motor neuron diseases such as amyotrophic lateral sclerosis (ALS). The clinical manifestations of ALS are diffuse and progressive muscle weakness of the extremities, bulbar paresis, and fatal respiratory failure caused by paresis of

both upper and lower motor neurons. CSA is clinically characterized by limited segmental muscle atrophy and corresponding multisegmental neurogenic EMG changes, but these changes are not as diffuse as those of ALS. However, differentiation of CSA from ALS, especially in the early stage of ALS, is difficult.¹¹⁾ Careful neurological, neuroradiological, and neurophysiological assessments are mandatory to differentiate CSA from ALS, before surgical treatment for CSA is planned.¹⁵⁾

The treatment for CSA is still controversial. Conservative treatment such as neck traction may be effective but complete resolution cannot be expected.^{3,7)} Cervical anterior decompression or posterior decompression (laminoplasty with or without foraminotomy) procedures have been advocated.^{8,10,12)} Compressive pathological lesions causing CSA are most often located anterior to the spinal cord and nerve root, so can justify anterior decompressive surgery. Therefore, we consider that in clinically suspected CSA and concomitant radiographically verified compression, surgery for anterior decompression is indicated. In the present three cases, we performed anterior decompression surgery at the C4-5 and C5-6 spaces. However, the level of decompression should be carefully determined according to not only neuroradiological findings but also clinical symptoms including electrophysiologic data.

We performed anterior decompression with titanium cage fusion in our three patients. Skin incision contralateral to the affected side facilitated a better view of the compressing spur and disc. The vertebrotomies were intentionally deviated to the affected side by a few mm from the midline to allow easier drilling of the posterolateral osteophytes. Drilling of the bony spur under the operating microscope was especially focused on the paramedial to lateral area, but the necessity for further foraminotomy depended on the individual preoperative neuroimaging findings. Opening of the paramedial to lateral (preforaminal) area through the intentionally deviated vertebrotomy allowed effective decompression of the anterolateral part of the spinal cord and the ventral root. After confirmation of complete decompression of the paramedial to lateral dura of the spinal cord, the vertebral bodies were fused with a titanium cervical cage. Titanium cervical cages widen the intervertebral disc space and consequently widen the intervertebral foraminal space without graft collapse, and provide immediate strong anterior interbody fusion.¹⁸⁾ This mechanical effect was confirmed by postoperative cervical radiography, which showed more lordotic spinal alignment after titanium cage fixation.

Two of our three patients almost fully recovered muscle strength and one patient showed moderate recovery after surgery. All three patients suffered muscle atrophy in the shoulder girdle and upper extremity at the time of surgery but MR imaging showed no abnormal intramedullary intensity. The effectiveness of surgical intervention for cervical spondylotic disorders depends on patient age, various clinical and radiological findings such as the interval between onset of symptoms and treatment, severity of symptoms, and presence of muscle atrophy, and the presence of intra-spinal hyperintensity on MR imaging.^{2,14,16)} The interval between onset and treatment was less than 3 months in the patients who made a good recovery (Cases 1 and 2), but was 18 months in the patient who showed moderate improvement (Case 3). This interval may be one of the most important indicators to predict the surgical result of CSA. Muscle power recovered after the surgery, despite the persistent muscle atrophy, suggesting that functionally reversible nerve fibers and anterior horn motor nuclei are still present if the surgical intervention is performed in the early stage of CSA. Intramedullary hyperintensity on T₂-weighted MR imaging of cervical spondylosis, called the "snake-eye appearance,"⁶⁾ may reflect cavity formation in the anterior horn caused by CSA.³⁾ The presence of hyperintensity may represent nonreversible pathological change, such as cavitation or cystic necrosis in the spinal gray matter.⁵⁾ Whether patients with CSA and such an intramedullary lesion will improve after surgical intervention remains controversial.^{7,8)} Although long-term follow up is required, our surgical experiences suggest that anterior surgical intervention may be effective despite severe muscle atrophy unless MR imaging shows the presence of a severe degenerative intramedullary lesion, especially in the anterior horn.

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