Distal-type cervical spondylotic amyotrophy: incidence and outcome after central corpectomy

Clinical article

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Object. Distal-type cervical spondylotic amyotrophy (CSA) is a rare form of cervical spondylotic myelopathy (CSM). The authors documented the incidence, clinical presentation, radiological features, and outcome following central corpectomy (CC) in patients with this entity.

Methods. The authors performed a retrospective institutional database search of patients who underwent decompressive surgery for CSM between 1992 and 2006 to identify patients with distal-type CSA. Distal-type CSA was defined as weakness and wasting of hands and forearms without gait impairment (Nurick Grades 0 and 1) and or sensory symptoms or signs in the lower limbs.

Results. The authors identified 7 male patients (1.1%) with distal-type CSA from among 653 patients who underwent either cervical laminectomy (135 patients) or CC (518 patients). There were sensory symptoms or signs in the upper limbs in all but 1 of the patients. Increased signal intensity in the cord was demonstrated on T2-weighted MR images in all patients. The compression was mainly at the C-6 vertebral level. At a mean follow-up of 46.5 months (range 12–98 months), 6 patients had improved by a mean patient perceived outcome score of 66.7% (range 20–100%). Patients’ modified Japanese Orthopedic Association scores improved from a preoperative mean (± SD) of 16.1 ± 0.7, to a follow-up mean of 17.4 ± 0.5 (p = 0.004, paired t-test). One patient whose condition worsened 7 months after CC received a diagnosis of a coexistent motor neuron disease.

Conclusions. Distal-type CSA is a rare form of CSM that should be differentiated from motor neuron disease on the basis of subtle sensory symptoms or signs in the upper limbs, and the presence of significant cord compression on the MR imaging. Patient outcome after central corpectomy is good and long lasting.

Key Words • cervical spine • corpectomy • outcome • spinal cord

Involvement of hand function in patients with CSA is commonly due to pyramidal weakness (“myelopathy hand”) or posterior column involvement (“numb clumsy hand”). Occasionally, patients with CSA present with predominant wasting and weakness of the upper limbs with minimal or no sensory symptoms; this condition is referred to as CSA. The affected muscles may be predominantly proximal such as the deltoid, biceps, and triceps (proximal CSA), distal such as the small muscles of the hands and forearm (distal CSA) or both proximal and distal (diffuse CSA). Cervical spondylotic amyotrophy is an uncommon syndrome with most authors reporting a single patient or only small series of patients.

Abbreviations used in this paper: ALS = amyotrophic lateral sclerosis; CC = central corpectomy; CSA = cervical spondylotic amyotrophy; CSM = cervical spondylotic myelopathy; EMG = electromyography; mJOA = modified Japanese Orthopaedic Association; MND = motor neuron disease; OPLL = ossified posterior longitudinal ligament; PLL = posterior longitudinal ligament.

Most reports of CSA have come from Japan and have focused on the proximal type typically associated with weakness of the deltoid muscle. Only a few authors have reported the outcome in distal CSA. Distal CSA is less frequently encountered than the proximal type. To the best of our knowledge there are no data on the incidence of this entity relative to the more common clinical presentation in patients with CSM and OPLL. The long-term outcome in patients with distal CSA after ventral decompressive surgery has also not been documented. In this study, we report the clinical and radiological presentation, management, and outcome of distal CSA in 7 patients. To the best of our knowledge this is the first report of a series of patients with distal CSA from outside Japan.

Methods

We identified patients with CSA through a retrospective search of a database of 653 patients who underwent decompressive surgery CC (518 patients) or laminectomy
Cervical spondylotic amyotrophy

(135 patients) for CSM or OPLL between 1992 and 2006 at our institution. We diagnosed the disease as distal type CSA if the following criteria were fulfilled: 1) a Nurick grade of 0 or 1 (no gait impairment); and 2) symptoms and signs present only in the distal parts of the upper limbs.

Patients with predominant motor involvement of the hands and forearms but with a Nurick grade > 1 or involvement of posterior column sensations in the lower limbs were excluded from the study.

Imaging Studies

All patients underwent preoperative imaging consisting of dynamic lateral cervical spine radiographs and MR images of the cervical spine. Sagittal T2-weighted MR images were examined for increased signal intensity within the spinal cord. Immediate postoperative radiography of the cervical spine was performed to confirm the position of the graft. At follow-up, dynamic radiographs of the cervical spine were obtained to confirm graft fusion and look for any instability.

Surgical Procedure

All patients underwent surgery performed by a single surgeon (V.R.). None of the patients who underwent cervical laminectomy had distal CSA. All patients underwent uninstrumented CC using autologous iliac or fibular bone graft in a surgical procedure that has been described in a previous publication.19 The width of the corpectomy was 14–16 mm and the PLL was excised in all patients until the dural tube was seen to bulge into the corpectomy defect.

Functional Evaluation

Functional status was evaluated based on Nurick grade and mJOA score preoperatively and at the last follow-up examination. At follow-up, all patients were examined clinically and asked to quantify their improvement on a scale of 0–100 compared to their preoperative status.14 This patient perceived outcome measure has been validated using construct validity and scores > 50 were considered to indicate a good outcome, 1–50 a fair outcome, and 0 or negative, a poor outcome.

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Duration of Sx (mos)</th>
<th>UL Motor Segments Involved</th>
<th>Sensory Symptoms in ULs</th>
<th>Sensory Loss in ULs</th>
<th>DTRs in LLs</th>
<th>Plantar Reflexes</th>
<th>Sensory Loss in LLs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>48</td>
<td>12</td>
<td>rt C7–T1</td>
<td>yes</td>
<td>yes</td>
<td>normal</td>
<td>normal</td>
<td>none</td>
</tr>
<tr>
<td>2</td>
<td>34</td>
<td>5</td>
<td>bilat C7–T1; rt&gt;lt</td>
<td>yes</td>
<td>yes</td>
<td>exaggerated</td>
<td>normal</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>37</td>
<td>5</td>
<td>bilat C7–T1</td>
<td>yes</td>
<td>yes</td>
<td>normal</td>
<td>normal</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>55</td>
<td>6</td>
<td>lt C6–T1</td>
<td>no</td>
<td>yes</td>
<td>normal</td>
<td>normal</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>53</td>
<td>4</td>
<td>bilat C7–T1</td>
<td>yes</td>
<td>yes</td>
<td>exaggerated</td>
<td>upgoing</td>
<td>none</td>
</tr>
<tr>
<td>6</td>
<td>45</td>
<td>12</td>
<td>bilat C7–T1; rt&gt;lt</td>
<td>yes</td>
<td>no</td>
<td>exaggerated</td>
<td>upgoing</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
<td>53</td>
<td>24</td>
<td>bilat C7–T1; rt&gt;lt</td>
<td>no</td>
<td>no</td>
<td>normal</td>
<td>normal</td>
<td>none</td>
</tr>
</tbody>
</table>

* DTR = deep tendon reflex; LL = lower limb; UL = upper limb.

Results

We identified 7 patients (1.1%) with features of distal CSA. All the patients were men with a mean age of 46.4 years (range 34–55 years). The duration of symptoms was 4–24 months (mean 8.3 months).

Clinical Features

All patients had unilateral or bilateral weakness and wasting of intrinsic muscles of the hands (Table 1). In 3 patients, the symptoms were asymmetric involving the right upper limb. However, examination revealed involvement of both hands in 2 of these patients. Two patients also had wasting of the forearm muscles, and all patients had weakness of wrist and/or elbow extension.

All but 2 patients complained of paresthesias in the upper limbs involving mainly the hands and the forearm in 1 patient. All except 2 patients had sensory deficits involving mainly the C-6, C-7, and T-1 dermatomes. In 1 patient the C-5 dermatome was also involved. Sensory loss ranged from 10 to 30%. Touch and pain modalities were mostly impaired, with joint position sense and vibration sense being impaired in the fingers in 1 patient.

Three patients had pyramidal signs in the lower limbs in the form of exaggerated deep tendon reflexes, but plantars were extensor in only 2 of these patients. None of the patients complained of difficulty in walking or had sensory deficits in the lower limbs. One patient (Case 7) reported urinary urgency and frequency but had no features of spasticity in the lower limbs and therefore the bladder symptoms were considered to be nonneurogenic in origin. No patient complained of neck or radicular pain.

The patient in Case 7 initially presented with hand tremors, more on the right side than the left, both action and postural, that had been going on for a year. He had no hand muscle weakness, questionable wasting of the right interossei muscles, and no other neurological deficits. The neurologists made a diagnosis of essential tremor. The patient did not undergo an EMG study that was recommended at that time. He presented 14 months later with weakness and more obvious wasting of his intrinsic hand muscles; an EMG obtained at this visit revealed denervation of the muscles supplied by the C-8 and T-1 segments.
bilateral and MR images demonstrated significant cord compression. The patient had no features of myelopathy.

**Management and Outcome**

Based on assessments of the sagittal alignment of the cervical spine on preoperative radiographs, 2 patients had lordotic spines and the other 5 had either straight or kyphotic spines. Magnetic resonance imaging showed compression of the cervical cord predominantly behind the C-6 vertebral body and adjacent disc spaces in 6 patients and the C-5 level in 1 patient. Additional levels of compression were seen in 3 patients with C-6 level compression. There was increased signal intensity in the cord on T2-weighted images in all patients. In 3 patients the classic “snake eyes” appearance was seen (Fig. 1 and Table 2).

Central corpectomies were performed at C-6 in all but 1 patient, and 1 patient each underwent a C-5 CC, a C5–6 corpectomy, and a C5–7 CC. Ossified PLLs were seen in 2 patients and chondroid metaplasia of the PLL in 1 patient. The PLL was thickened in 3 other patients and appeared normal in 1. There was no evidence of herniation of the disc fragments either within the layers of the PLL or behind the PLL in any patient.

All 7 patients were available for follow-up. The mean follow-up period was 46.5 months (range 12–98 months) in the 6 patients whose condition improved. The progressive wasting of the hands had been arrested and hand muscle functioning had improved. The mean improvement on the patient perceived outcome score was 66.7% (range 20–100%). Four patients reported a good outcome, 2 reported a fair outcome, and 1 patient reported worsening. The patient in Case 6 reported worsening of symptoms 7 months after surgery. An EMG study obtained at that time revealed denervation in the bulbar muscles and a diagnosis of MND was rendered. Repeated MR imaging was done to rule out further compression (Fig. 2). The Nurick grade improved in 1 patient and remained the same in the other 6. There was a significant improvement in the mJOA scores from a preoperative mean of 16.1 ± 0.7 to a mean score at follow-up of 17.4 ± 0.5 (p = 0.004, paired t-test; 95% CI −1.98 to −0.59)

**Discussion**

**Clinical Features of CSA**

Cervical spondylotic amyotrophy is an atypical presentation of CSM that was initially described by Brain and Wilkinson. However, it was Yanagi et al. in 1976 who introduced the term CSA. The motor segments involved in patients with this condition are C7–8 and T-1, either unilaterally or bilaterally. In patients with bilateral involvement there is considerable asymmetry, a feature we noted in our patients. Although most of our patients complained of symptoms in 1 upper limb, careful neurological examination revealed mild deterioration of muscle power in the other limb in 4 of our patients. The sensory dysfunction in patients with CSA is mild with slight hypesthesia in the upper limbs. Pyramidal tract involvement is usually limited to exaggerated deep tendon reflexes in the lower limbs without gait disturbance. This reflects the predominant involvement of the central gray matter of the cervical cord with minimal involvement of the long tracts. However, the clinical features of patients with distal CSA as reported in the literature have varied considerably. Some authors have included patients with posterior column involvement in the lower limbs and/or severe gait disturbance.2 Our patients were more homogenous in their presentation; none had gait impairment or sensory deficits in the lower limbs. Our patients therefore fit the definition of CSA proposed by Ebara and colleagues.3 A clear definition of this clinical entity is important so that confusion with the more common presentation of CSM is avoided. Patients with CSM present mainly with gait impairment and with variable degrees of upper limb involvement. The clinical features also have an important bearing on our understanding of the causes of this disease.

Patients with distal CSA rarely experience progression of the disease to the proximal muscles.20 None of our patients had any wasting of the proximal muscles. It has been reported that patients with distal CSA have a longer duration of symptoms than those with the proximal type; 3 of our patients had symptoms for a year or more.

**Pathophysiological Characteristics of CSA**

It has been suggested that patients with CSA have either compression of the ventral roots or the anterior horn by bone spurs.10 Ischemic damage to these structures as a result of the compression has been suggested to be the main cause of the disease.8,21 It is unlikely that the compression of 1 or 2 radicular arteries can cause in-
volvement of a significant enough segment of the anterior horn to cause severe muscle wasting, as most upper limb muscles are innervated by > 1 segment. It is more likely that the hypoperfusion in the distribution of the anterior spinal artery is the cause of the ischemia of a significant longitudinal extent of the ventral horn resulting in a multisegemental damage to the cord, and wasting and weakness of the muscles. Tsuboi et al. suggest that venous congestion is the cause of the gray matter involvement of the cervical cord. Ebara et al. reported postmortem findings of severe infarction of the gray matter with mild damage to the pyramidal tract in the cervical cord of a patient with distal CSA. However, all the findings in patients with CSA cannot be explained solely by the involvement of the gray matter. The presence of sensory symptoms in the upper limbs indicates involvement of the spinal root in the root canal rather than the ventral motor root alone, and the presence of pyramidal signs in the lower limbs indicates damage to the lateral corticospinal tracts. Although patients with proximal CSA could have either root or anterior horn compression, those with the distal type are more likely to have anterior horn compression with or without additional root compression. Multisegmental involvement is also more often noted in patients with distal CSA than in those with the proximal type. We believe that cord involvement in our patients was the chief cause of the deficits, as shown by the clinical and radiological features.

Intraoperative spinal cord evoked sensory potentials in 6 patients with distal CSA revealed attenuation of the N13 peak with preserved N9 and N11 peaks indicating a postsynaptic block in the gray matter rather than at the level of the roots. Intraoperative motor evoked potentials in the same group of patients revealed abnormalities in 4 patients with clinical features of pyramidal tract involvement. Fujiwara et al. also found conduction blocks in the spinal tracts even in patients without clinical features of spinal tract involvement.

**Differential Diagnosis**

Distal CSA must be distinguished from anterior horn cell diseases such as ALS. The presence of subtle but definite sensory signs or symptoms usually reveals the diagnosis, and bulbar muscle involvement obviously points to the diagnosis of ALS. Magnetic resonance imaging will also reveal cord changes in patients with CSA. The difficulty arises when the patient has both diseases or when MR imaging in a patient with MND shows mild cord compression. One of our patients had both disease processes, and there are reports of similar cases in literature. We suggest that surgery only be offered to patients with evidence of definite cord compression in the form of intramedullary cord changes on T2-weighted MR images. Our patient with dual pathology (Case 6) had definite cord compression on the MR with increased signal inten-

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**TABLE 2: Summary of imaging features, treatment, and outcome in patients with distal CSA**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Level of Compression on MRI</th>
<th>Intramedullary Signal Changes</th>
<th>Operative Findings</th>
<th>Surgery</th>
<th>PPOS (FU in mos)</th>
<th>Nurick Grade Preop/Postop</th>
<th>mJOA Score Preop/Postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>C5–7</td>
<td>yes (snake eyes)</td>
<td>OP LL</td>
<td>C5–7 CC &amp; fibular graft</td>
<td>75% improved (45)</td>
<td>0/0</td>
<td>17/18</td>
</tr>
<tr>
<td>2</td>
<td>C-6</td>
<td>yes</td>
<td>thick PLL</td>
<td>C-6 CC &amp; iliac graft</td>
<td>60% improved (34)</td>
<td>1/0</td>
<td>16/18</td>
</tr>
<tr>
<td>3</td>
<td>C-6</td>
<td>yes (snake eyes)</td>
<td>thick PLL</td>
<td>C-6 CC &amp; iliac graft</td>
<td>95% improved (72)</td>
<td>1/1</td>
<td>16/17</td>
</tr>
<tr>
<td>4</td>
<td>C-6</td>
<td>yes (snake eyes)</td>
<td>chondroid metaplasia of PLL</td>
<td>C-6 CC &amp; iliac graft</td>
<td>20% improved (12)</td>
<td>1/1</td>
<td>15/17</td>
</tr>
<tr>
<td>5</td>
<td>C-5</td>
<td>yes</td>
<td>normal PLL</td>
<td>C-5 CC &amp; iliac graft</td>
<td>100% improved (98)</td>
<td>1/1</td>
<td>16/18</td>
</tr>
<tr>
<td>6</td>
<td>C5–6</td>
<td>yes</td>
<td>thick PLL</td>
<td>C5–6 CC &amp; iliac graft</td>
<td>worse (7)</td>
<td>1/1</td>
<td>17/17</td>
</tr>
<tr>
<td>7</td>
<td>C5–6</td>
<td>yes</td>
<td>OPLL</td>
<td>C5–6 CC &amp; iliac graft</td>
<td>50% improved (18)</td>
<td>0/0</td>
<td>16/17</td>
</tr>
</tbody>
</table>

* FU = follow-up; PPOS = patient perceived outcome score.
sity within the cord. Shindo et al. have found that muscle sympathetic nerve activity was significantly elevated in patients with ALS and cervical spondylisis compared with those with CSA. Other authors have also noted coexistence of CSA with another pathological entity such as MND. Dorsen and Ehni reported 2 such cases: worsening of symptoms in the postoperative period developed in 1 patient who later received a diagnosis of MND. Another of their patients had features of MND as well as spondylotic compressive elements and did not show any postoperative improvement. One of the 7 patients who underwent surgery in the study of Ebara et al. did not show postoperative improvement, and they suspected that he had a dual pathology because of the discrepancy between the location of the cord compression on imaging and the clinically evident involved motor segments.

Shibuya and colleagues suggest that some patients with an anterior spinal artery syndrome, might be misdiagnosed as having CSA. One of the key features to differentiate the 2 is the acute presentation of anterior spinal artery syndrome in contrast to the more slowly progressive clinical symptoms in patients with CSA.

**Imaging Studies**

Several authors have documented increased signal intensity in the cord in patients with distal CSA. Typical changes have involved the anterior horn region or the gray matter as a whole, and these have been characterized as “snake eyes” changes. Because all of our patients had intramedullary high-intensity signal changes on T2-weighted MR images, it can be presumed that in our patients the cause of amyotrophy was damage to the anterior horns of the cord rather than to the ventral roots. Fujiwara reported on a patient with distal CSA in whom delayed CT myelography revealed cavitation within the anterior horn on the affected side. The authors concluded that this finding clearly points to the involvement of the gray matter in patients with CSA.

Tsuboi et al. found cord compression mainly at the C5–6 or the C6–7 intervertebral disc level in their patients with CSA. In 3 of our patients the compression was exclusively behind the C5–6 and C6–7 disc levels while in 3 others the compression included other adjacent levels. In only 1 patient was the compression restricted to the C4–5 and C5–6 disc levels.

**Management Options**

Almost all Japanese authors who reported on patients with distal CSA resorted to laminoplasty as the treatment of choice (Table 3). Conservative therapy seems to arrest disease progression in some patients with proximal CSA, but has not been documented to be effective in patients with distal CSA. Ebara et al. suggested that patients with distal CSA undergo cervical traction for an unspecified period of time and that they should only undergo surgery if there was improvement in their symptoms. These authors did not, however, elaborate on the rationale for following this line of management. Ebara et al. also preferred laminoplasty to CC because the disease was in the cord and not in the roots. Again their rationale for this is unclear as the compression is definitely ventral to the cord in most patients with distal CSA. Therefore, we consider a ventral decompressive procedure such as CC to be ideal. Our bias is toward ventral decompression in all patients with CSM as the compression is anterior in most patients. Furthermore, 5 of the 7 patients in our series had a straight or kyphotic curvature of the cervical spine and therefore, would not have been ideal candidates for posterior decompression. We prefer corpectomy to discectomy whenever compression is present behind ≥ 2 adjacent disc spaces and when the disease extends behind the bodies as in the patients with OPLL.

The outcome after surgical decompression is superior in patients with proximal CSA than in those with the distal variant. Fujiwara et al. reported that the muscle power improved in 92% of their patients with the proximal CSA but in only in 38% of those with distal CSA. A possible reason for this might be the fact that proximal CSA can be either in the roots or in the anterior horns, whereas patients with the distal type uniformly have involvement of the anterior horns. The average duration of symptoms is also longer in patients with distal CSA, which might again explain the worse outcome in these patients compared to those with the proximal type. The formal testing of the motor power of the hand muscles in patients with distal CSA might not show any improve-

**TABLE 3: Summary of literature on treatment and outcome in patients with distal CSA**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Duration of Sx (mos)</th>
<th>Sensory Loss in LLs</th>
<th>Gait Impairment</th>
<th>No. of Patients W/ ISI on MRI</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ebara et al., 1988</td>
<td>7</td>
<td>ND</td>
<td>no</td>
<td>no</td>
<td>not done</td>
<td>laminoplasty (6 patients); CC (1 patient)</td>
<td>GS improved in 6</td>
</tr>
<tr>
<td>Kaneko et al., 2004</td>
<td>6</td>
<td>ND</td>
<td>yes (mild)</td>
<td>yes (4 patients)</td>
<td>6</td>
<td>laminoplasty</td>
<td>GS improved in 4</td>
</tr>
<tr>
<td>Fujiwara et al., 2006</td>
<td>8</td>
<td>3–60 (mean 12.3)</td>
<td>no</td>
<td>no</td>
<td>6</td>
<td>laminoplasty &amp; foraminotomy</td>
<td>5 had no change in MP; 3 improved by 1 grade MP</td>
</tr>
<tr>
<td>present study</td>
<td>7</td>
<td>4–24 (mean 8.3)</td>
<td>no</td>
<td>no</td>
<td>7</td>
<td>CC</td>
<td>6 improved; 1 worsened (had MND)</td>
</tr>
</tbody>
</table>

* GS = grip strength; ISI = increased signal intensity; MP = motor power; ND = no data.
Cervical spondylotic amyotrophy

ment after surgery. We found that hand function definitely improves postoperatively, as shown in the present study in which 6 of our 7 patients, self-reported improvement ranged from 20 to 100%.

Conclusions

Distal CSA is a rare presentation in patients with CSM or OPLL, and must be distinguished from MND although rarely the conditions can coexist. Most often the pathological entity is in the cord and not in the roots, with the compression present at the C-6 vertebral level. Central corpectomy provides long lasting improvement in symptoms of patients with distal CSA.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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