Cervical Myelopathy Caused by Hypoplasia of the Atlas and Ossification of the Transverse Ligament

—Case Report—

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Abstract

A 79-year-old Japanese female presented with symptomatic cervical myelopathy caused by a hypoplastic posterior arch of the atlas and ossification of the transverse ligament. Neuroradiological examination demonstrated a hypoplastic posterior arch of the atlas and ossification of the transverse ligament. The cervical spinal cord was compressed at the level of the atlas by both the hypoplastic posterior arch of the atlas and the ossification of the transverse ligament. The patient underwent C-1 laminectomy, which arrested the progressive myelopathy and resulted in a good recovery. Atlas hypoplasia with ossification of the transverse ligament may be associated with Asian ethnicity.

Key words: atlas, hypoplasia, transverse ligament, myelopathy, ossification

Introduction

Congenital anomalies of the posterior arch of the atlas (C-1) are uncommon. Non-traumatic cervical myelopathy caused by canal stenosis at the level of the atlas is extremely rare, with only 12 reported cases.9,11–14,16,18,21,24,27) Ossification of the vertebral ligaments, most commonly the posterior longitudinal ligament (OPLL), frequently occurs in the Japanese population.15) However, symptomatic ossification of the transverse ligament is extremely rare.1,6) Here, we describe a case that involved both congenital stenosis at the level of the atlas, caused by a hypoplastic posterior arch of the atlas, and ossification of the transverse ligament.

Case Report

A 79-year-old Japanese female presented with a one-year history of occipitalgia. Her gait had been impaired for several months, requiring the use of a cane. She had no history of head or neck trauma. Routine laboratory tests, including serum calcium, were normal. Neurological examination on admission revealed that the cranial nerves were normal. The range of motion of the head was normal, and the patient showed no torticollis. The patient had right hemiparesis and loss of sensation in both arms. She showed hyperreflexia in all four extremities with positive bilateral Babinski’s and Hoffmann’s signs.

Cervical radiography demonstrated a markedly anterior position of the posterior arch of the atlas with reduced spinal canal diameter (Fig. 1). No instability was detected. Magnetic resonance imaging revealed spinal cord compression at the level of the atlas and the C3–4 vertebrae (Fig. 2). Computed tomography (CT) myelography demonstrated marked narrowing of the spinal canal and ossification of the transverse ligament. The anterior-posterior diameter of the spinal canal was only 8 mm, and both the subarachnoid space and the spinal cord were compressed (Fig. 3 left). Therefore, the major cause of the cervical myelopathy was thought to be atlas hypoplasia and concomitant ossification of the transverse ligament.
Fig. 1 Radiographs of the cervical spine, lateral view, showing marked stenosis of the spinal canal at the level of the atlas as well as degenerative changes below C-1. A: flexion, B: neutral, C: extension.

Fig. 2 Sagittal T₁-weighted (left) and T₂-weighted (right) magnetic resonance images showing the hypoplastic posterior arch of the atlas has reduced the cervical canal diameter, resulting in cord compression.

Fig. 3 Preoperative computed tomography (CT) myelogram demonstrating a narrow spinal canal at the level of the atlas and an ossified transverse ligament (left). Postoperative CT myelogram showing the decompressed spinal cord (right).

The patient underwent laminectomy of the atlas via a posterior approach. The small posterior arch of C-1 was identified and was resected to achieve complete decompression. Postoperative CT myelography confirmed decompression of the spinal cord at the C-1 level (Fig. 3 right). The postoperative course was uneventful. Her occipitalgia disappeared, and the right hemiparesis improved. The patient was able to walk without a cane. The patient’s condition has been stable for 18 months after surgery.

Discussion

This case showed an extremely unusual association of atlas hypoplasia with ossification of the transverse ligament resulting in cervical myelopathy. The
spinal cord was compressed by both the ossified transverse ligament and the hypoplastic posterior arch of the atlas.

The majority of anomalies of the posterior arch of the atlas consist of various arch clefts, aplasias, and hypoplasias, which can best be understood by reference to the atlas ossification centers. The majority of the atlas originates in the rostral half of the first cervical sclerotome, and the atlas has lateral and anterior ossification centers. Chondrification of the posterior arch begins at the pedicles during the 6th week of embryogenesis and ends at the midline during the 4th month. This cartilaginous arch ossifies by 3 to 4 years of age. Five cases of cervical myelopathy have been caused by spina bifida of the posterior arch of the atlas in children, in which the concave halves of the arch of the atlas compressed the spinal cord. The cause of the bifid posterior arch may be either failure of the extension process of the chondrification centers in the posterior arch, or failure of the ossification process. In contrast, our patient had a complete posterior arch of the atlas resulting in a narrow spinal canal, which suggests that this particular anomaly may have a different embryological origin, such as premature fusion of cartilaginous synchondrosis or abnormal rotary biomechanics causing facial hypertrophy.

The transverse ligament is about 20 mm long and 2 mm thick but becomes thinner toward the center. This ligament is involved in the stabilization of the atlantoaxial joint by supporting the dens together with the cruciform ligament. Ossification of the transverse ligament is extremely rare. Only one of more than 1000 postmortem spinal specimens showed calcification of the transverse ligament of the atlas. Only six cases of ossification of the transverse ligament have been reported, and of these only two cases showed myelopathy due to spinal cord compression caused by the ossified transverse ligament. Calcification and ossification are difficult to discriminate without histological specimens. Ossification usually originates from the rim of the ligament. In contrast, calcification originates from the center of the ligament. In our case, CT myelography clearly demonstrated that the degenerative change originated from the rim of the ligament (Fig. 3). Although removal or biopsy of the ligament was not performed, we considered this degenerative change as ossification.

The factors involved in ossification of the transverse ligament may include calcium-phosphate metabolic disease, diabetes mellitus, obesity, and aging, but no consensus has been achieved. The cause of ossification of the transverse ligament in our case may have been degeneration as a result of aging or ankylosing process by dynamic factors such as trauma. However, there may be an accessory anterior ossification center that can cause the ossification of the transverse ligament in addition to the anterior and lateral ossification centers. Our case also had atlas hypoplasia, a developmental anomaly that may be related to the presence of an accessory ossification center.

Only 12 cases of non-traumatic symptomatic cervical myelopathy caused by atlas hypoplasia have been reported (Table 1). Interestingly, all 13 known cases, including the present case, occurred in individuals of Asian origin (10 Japanese, 2 Chinese, 1 unknown). OPLL commonly occurs in the Japanese population, so the ethnic association is very important for this pathology. Patients with OPLL tend to have hyperostosis of the spinal ligaments, as ankylosing spinal hyperostosis has been observed in 23.9% to 30% of patients with OPLL. One case of atlas hypoplasia was associated with ankylosing spinal hyperostosis (Case 4, Table 1). Therefore, we can conclude that these two clinical entities show similarities. An ethnic association for this anomaly was suggested by an analysis of five cases. Here, we have included eight more cases of the phenomenon, which strongly suggest that there is an ethnic association for this type of anomaly.

The normal retrodental space is 17 to 25 mm at the level of the atlas, whereas the spinal cord diameter ranges from 10 to 12 mm. This difference explains the lower incidence of symptomatic canal stenoses at the level of the atlas. Spinal cord compression should be considered a possibility if the sagittal canal diameter is less than 14 mm. The 13 known cases had a retrodental space at the level of the atlas of between 7 and 11 mm (mean 8.3 ± 1.4 mm). However, the clinical manifestations occurred relatively late in life (mean 66.3 ± 13.0 yrs). The youngest patient was a 38-year-old male (Case 1) who also had Chiari malformation, so the herniated cerebellar tissue extending into the foramen magnum would influence the symptoms. All other cases were complicated by canal stenosis below the atlas (Cases 1, 4, 8, and 9), spondylotic change (Cases 2, 4–9, 11, and 12), OPLL (Case 10), or retroodontoid soft tissue mass (Cases 4, 6, and 7). Our case (Case 13) was complicated by ossification of the transverse ligament. Furthermore, most patients had suffered from long-standing myelopathic symptoms (mean 46.5 ± 5.7 yrs). Therefore, the presence of a congenital canal stenosis at the level of the atlas became symptomatic when complicated by chronic degenerative changes such as spondylotic change, ligament hypertrophy or ossification, or injury due to hyperextension.
Table 1 Summary of patients with atlas hypoplasia

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/ Sex</th>
<th>Ethnicity</th>
<th>Symptoms (duration)</th>
<th>C-1 diameter (mm)</th>
<th>Other radiological findings</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Ishii et al. (1984)</td>
<td>55/M</td>
<td>Japanese</td>
<td>tetraparesis (10 yrs), sensory loss in all limbs</td>
<td>7</td>
<td>canal stenosis</td>
<td>C-1 laminectomy, C3–6 laminoplasty</td>
<td>MD</td>
</tr>
<tr>
<td>2</td>
<td>58/M</td>
<td>Japanese</td>
<td>sensory loss in all limbs (1 yr)</td>
<td>8</td>
<td>spondylotic change</td>
<td>C-1 laminectomy</td>
<td>MD</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Sawada et al. (1989)</td>
<td>38/M</td>
<td>Japanese</td>
<td>tetraparesis (3 yrs), sensory loss in all limbs</td>
<td>7</td>
<td>Chiari malformation</td>
<td>C-1 laminectomy</td>
<td>MD</td>
</tr>
<tr>
<td>4</td>
<td>Komatsu et al. (1993)</td>
<td>56/M</td>
<td>Japanese</td>
<td>tetraparesis (11 yrs), gait disturbance</td>
<td>7.7</td>
<td>canal stenosis, retroodontoid mass</td>
<td>occipital decompression, C-1 laminectomy, occipital decompression</td>
<td>MD</td>
</tr>
<tr>
<td>5</td>
<td>Tokiyoshi et al. (1994)</td>
<td>55/M</td>
<td>Japanese</td>
<td>tetraparesis (4 mos), sensory loss in both arms</td>
<td>8</td>
<td>spondylotic change</td>
<td>C-1 laminectomy, occipital decompression</td>
<td>GR</td>
</tr>
<tr>
<td>6</td>
<td>Kubo et al. (1995)</td>
<td>69/M</td>
<td>Japanese</td>
<td>tetraparesis (3 yrs), sensory loss in both arms</td>
<td>10</td>
<td>spondylotic change, retroodontoid mass</td>
<td>C-1 laminectomy, posterior fusion</td>
<td>MD</td>
</tr>
<tr>
<td>7</td>
<td>Yamashita et al. (1997)</td>
<td>73/F</td>
<td>Japanese</td>
<td>tetraparesis (3 yrs), sensory loss in both arms</td>
<td>11</td>
<td>spondylotic change, retroodontoid mass</td>
<td>C-1 laminectomy, posterior fusion</td>
<td>GR</td>
</tr>
<tr>
<td>8</td>
<td>Noguchi et al. (1998)</td>
<td>81/M</td>
<td>Japanese</td>
<td>tetraparesis (3 yrs), sensory loss in all limbs</td>
<td>10</td>
<td>canal stenosis</td>
<td>C-1 laminectomy, occipital decompression</td>
<td>MD</td>
</tr>
<tr>
<td>9</td>
<td>Phan et al. (1998)</td>
<td>80/M</td>
<td>Chinese</td>
<td>tetraparesis (2.5 yrs), sensory loss in both arms</td>
<td>8</td>
<td>canal stenosis</td>
<td>C-1 laminectomy, occipital decompression</td>
<td>MD</td>
</tr>
<tr>
<td>10</td>
<td>75/M</td>
<td>Chinese</td>
<td>tetraparesis (1.5 yrs), sensory loss in all limbs</td>
<td>7</td>
<td>C2–3 OPLL</td>
<td>C-1 laminectomy</td>
<td>MD</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Okamoto et al. (1998)</td>
<td>77/?</td>
<td>Japanese</td>
<td>spasticity (20 yrs)</td>
<td>11</td>
<td>spondylotic change</td>
<td>C-1 laminectomy, posterior fusion</td>
<td>GR</td>
</tr>
<tr>
<td>12</td>
<td>May et al. (2001)</td>
<td>66/M</td>
<td>?</td>
<td>rt upper limb numbness (1 yr)</td>
<td>10</td>
<td>spondylotic change</td>
<td>C-1 laminectomy, occipital decompression</td>
<td>MD</td>
</tr>
<tr>
<td>13</td>
<td>Present case</td>
<td>79/F</td>
<td>Japanese</td>
<td>hemiparesis (1 yr), sensory loss in both arms</td>
<td>7</td>
<td>spondylotic change, ossified transverse ligament</td>
<td>C-1 laminectomy</td>
<td>GR</td>
</tr>
</tbody>
</table>

GR: good recovery, MD: moderate disability, OPLL: ossification of the posterior longitudinal ligament.

spine, due to degenerative change, may also result in increased movement of the atlas with subsequent spinal cord compression.\(^{12,18}\) All patients underwent surgical decompression and had a favorable outcome (Table 1). Therefore, surgical removal of the hypoplastic posterior arch of the atlas may be the treatment of choice.

The present case of myelopathy caused by atlas hypoplasia associated with ossification of the transverse ligament in a Japanese female is of particular interest, because this anomaly is a phenomenon that is more common in Asians. Whether this finding indicates a real ethnic difference in the disease incidence or simply reflects the difference in attention toward this clinical entity remains unknown. Study of more patients with this condition will provide more evidence of the relationship between ethnic background and atlas hypoplasia, and ossification of the transverse ligament.

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