Atlas Hypoplasia Associated With Non-traumatic Retro-odontoid Mass

—Case Report—

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Abstract

A 38-year-old man presented with progressive cervical myelopathy due to atlas hypoplasia associated with non-traumatic retro-odontoid mass. The neuroimaging findings suggested hypertrophy of the transverse ligament of the atlas. No histological confirmation of the retro-odontoid mass was obtained. Clinical manifestations improved after posterior decompression. Decompressive laminectomy of the atlas with or without fusion can achieve a good outcome in such cases.

Key words: atlas hypoplasia, retro-odontoid mass, myelopathy, transverse ligament, laminectomy

Introduction

Non-traumatic cervical myelopathy caused by anomaly of the atlas is uncommon. Atlas hypoplasia is a quite rare congenital anomaly caused by dorsal expansion failure of two lateral primary ossification centers. Only 12 cases of atlas hypoplasia have been reported. On the other hand, various types of non-traumatic retro-odontoid extradural mass lesions may cause compression of the spinal cord, including rheumatoid pannus, calcium pyrophosphate dihydrate (CPPD) deposition disease, ossification of the transverse ligament, herniated disc, and synovial cyst. Only three cases combining atlas hypoplasia and non-traumatic retro-odontoid soft tissue mass have been reported. Here, we describe another case of myelopathy caused by both congenital atlas hypoplasia and acquired retro-odontoid mass lesion.

Case Report

A 38-year-old man was admitted to our hospital with a 3-month history of sensory and motor disturbance of the extremities. On admission, neurological examination demonstrated slight tetraparesis (left > right) with spastic gait, left hemihyperesthesia, exaggerated bilateral deep tendon reflexes, and left Babinski sign. There was no evidence of previous trauma or rheumatoid arthritis.

Lateral cervical spine radiography showed a

Fig. 1 Computed tomography scans showing marked canal stenosis due to the hypoplastic atlas and punctate mineralization beside the left lateral mass of the atlas.
hypoplastic atlas. There was no evidence of atlantoaxial dislocation or instability on flexion/extension. Computed tomography (CT) demonstrated marked canal stenosis at the level of the atlas, and the sagittal canal diameter was 12 mm (Fig. 1). Small punctate mineralization was also detected in the canal adjacent to the left lateral mass of the atlas. Sagittal magnetic resonance (MR) imaging demonstrated narrowing of the spinal canal at the level of the atlas, and T2-weighted MR imaging showed a cord signal intensity change which was interpreted as ischemic damage (Fig. 2A). Mild spondylosis was also seen from the C-3 to C-7 levels (Fig. 2A). Axial MR imaging demonstrated an extradural retro-odontoid soft tissue mass which severely compressed the spinal cord posteriorly, in addition to the hypoplastic atlas (Fig. 2B, C). T1-weighted MR imaging showed the retro-odontoid mass as intermediate to low intensity (Fig. 2B) and T2-weighted imaging as predominantly low intensity with partial high intensity (Fig. 2C). Both T1- and T2-weighted MR imaging demonstrated a curvilinear signal void between the spinal cord and retro-odontoid mass caused by the tectorial membrane17 (Fig. 2B, C). The space available for the spinal cord at the level of the atlas was 6 mm.

Decompressive laminectomy of the atlas was performed. The postoperative course was uneventful, and the patient’s tetraparesis and hyperesthesia gradually improved. MR imaging 6 years after the surgery showed marked decompression of the spinal cord, and decreased intramedullary signal change at the level of the atlas (Fig. 3). There was no remarkable change in the size of the retro-odontoid mass. However, spondylotic change from the C-3 to C-7 levels had progressed.

Discussion

The previous three and present cases of atlas hypoplasia associated with acquired non-traumatic retro-odontoid mass lesion causing spinal cord compression are summarized in Table 1.3,13,15 The four patients, two men and two women, were aged from 38 to 79 years (mean 61.5 years). Interestingly, all four cases occurred in Japanese patients, suggesting an ethnic and/or genetic factor in the development of this condition.
Table 1  Summary of cases of symptomatic atlas hypoplasia associated with non-traumatic retro-odontoid mass

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age (yrs)/Sex</th>
<th>Diameter of spinal canal/spinal cord (mm)</th>
<th>Radiological diagnosis of the retro-odontoid mass</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Komatsu et al. (1993)</td>
<td>56/M</td>
<td>7.7/—</td>
<td>soft tissue mass</td>
<td>C-1 laminectomy and occipital decompression</td>
<td>excellent</td>
</tr>
<tr>
<td>Yamashita et al. (1997)</td>
<td>73/F</td>
<td>11/—</td>
<td>pseudotumor</td>
<td>C-1 laminectomy and posterior fusion</td>
<td>excellent</td>
</tr>
<tr>
<td>Tsuruta et al. (2003)</td>
<td>79/F</td>
<td>7/—</td>
<td>ossification of the transverse ligament</td>
<td>C-1 laminectomy</td>
<td>excellent</td>
</tr>
<tr>
<td>Present case</td>
<td>38/M</td>
<td>12/6</td>
<td>hypertrophy of the retro-odontoid soft tissue</td>
<td>C-1 laminectomy</td>
<td>excellent</td>
</tr>
</tbody>
</table>

of these lesions. The most common symptoms were upper cervical myelopathy, which usually appeared during adulthood. The retro-odontoid masses were described as thickening of the soft tissue,3) pseudotumor,15) and ossification of the transverse ligament,13) all of which were identified by radiographic findings only. This type of acquired retro-odontoid mass lesion is one of the reasons for the late appearance of clinical symptoms, because the congenital hypoplastic atlas usually does not result in serious neurological signs.3,8,13–15)

The normal sagittal canal diameter at the level of the atlas is 16 to 20 mm in Asians.8,10,12) If the canal is less than 10 mm, clinical manifestations usually occur.3,4,10,12) In the present case, the diameters of the spinal canal and the spinal cord at the level of the atlas were 12 mm and 6 mm, respectively. Chronic degenerative changes causing late occurrence of clinical manifestations have also been identified, such as spondylotic change of the lower cervical spine.4,8,12,13,15) In our case, mild spondylotic change from C-3 to C-7 also complicated the patient’s condition. The decreased range of motion at the lower cervical levels from spondylosis may have resulted in increased movement at the level of the atlas, subsequently compressing the spinal cord.8)

MR imaging of the retro-odontoid mass in our case showed a striking similarity to pseudotumor,15) which is a fibrous granulation or fibrocartilaginous mass usually associated with chronic osteoarthrosis10) or atlanto-axial subluxation.11) This type of lesion appears as intermediate to low intensity on T1-weighted images and mainly low intensity on T2-weighted images, consistent with the fibrous composition.11,15) Other types of retro-odontoid lesions causing myelopathy include rheumatoid pannus,10) CPPD deposition disease,1,17) ossification of the transverse ligament,7,13) herniated disc,9) and synovial cyst.2) Our patient denied any history of trauma, and there was no evidence of rheumatoid arthritis. CT of the lesion in CPPD deposition disease shows heterogeneous calcification,1,17) which apparently differs from the findings in our case. Therefore, the radiological diagnosis of the retro-odontoid mass in our case was hypertrophy of the soft tissue including the transverse ligament due to chronic degeneration or inflammation of unknown etiology, despite the absence of dislocation. Calcification of the transverse ligament is a quite common degenerative change seen in atlanto-odontoid joint neck disease.16) However, ossification of the transverse ligament is rarer. In our case, although slight mineralization was detected in the retro-odontoid region, calcification and ossification were difficult to discriminate without histological examination.

The outcome in our patient and previous patients has been excellent after C-1 laminectomy with or without posterior fixation (Table 1).3,13,15) A case of pseudotumor treated by laminectomy of the atlas with occipitocervical fusion showed subsequent spontaneous reduction of the mass, probably due to stabilization of atlanto-axial joint.15) Therefore, additional posterior fusion is recommended, especially in cases complicated by severe osteoarthrosis. There were no complications in the present or previous cases. Although no long-term follow-up review is available, we suggest that decompressive laminectomy of the atlas is safe and effective treatment for atlas hypoplasia even if complicated by retro-odontoid mass.

References

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