Demonstration of craniocervical junction abnormalities for diagnosis of atlanto-occipital assimilation using MRI

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Abstract

Objectives: Atlanto-occipital assimilation (AOA) is one of the most common skeletal anomalies of the craniovertebral junction (CVJ). Because its clinical symptomatology is non-specific and it has several variations, many cases go unnoticed which may lead to additional and unnecessary radiological examinations. In this study, we aimed to present CVJ abnormalities with MRI to improve diagnostic accuracy of AOA.

Methods: Cervical MRIs of the patients registered in PACS between January 2008 and October 2011 were scanned and AOA was detected in 40 cases. Sagittal FSE T1 and T2-weighted cervical MRIs and axial T2*-GRE sequence images were re-evaluated for AOA typing, anterior atlantodental interval (AADI), posterior atlantodental interval (PADI) measurements, spine fusion anomalies, basilar invagination, tonsillar herniation, myelomalacia, suboccipital muscles and vertebral arteries (VAs).

Results: CVJ abnormalities were present in all cases and the most frequent association was observed in suboccipital muscles (100%) and VAs (95%). 60% of the cases had decreased PADI, 32% C2–3 vertebrae fusion, 25% increased AADI, 22.5% basilar invagination, 15% myelomalacia and 5% tonsillar herniation.

Conclusion: Suboccipital muscle abnormality was found in all AOA cases whatever the severity and type of the bony fusion. VA anomaly was observed as the second most common abnormality and accompanied preferably the cases with lateral body involvement. Being aware of additional CVJ abnormalities in MRI examinations may reduce unnecessary radiological examinations by increasing the AOA diagnosis rate.

Keywords: assimilation; craniocervical; magnetic resonance imaging

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malities which lead to weakness and ataxia in the lower extremities and numbness and pain in the upper limbs. Preoperative angiogram is strongly recommended for the demonstration of the course of the VA.\textsuperscript{\textcircled{1}}

Considering its symptomatology and prevalence, AOA may be overlooked and this may lead to unnecessary radiological examinations of different anatomical regions. As well as unnecessary costs caused by the delays in diagnosis, surgical procedures scheduled in patients who did not get a diagnosis may also pose risks for this patient group. AOA diagnosis is of vital importance for the radiologists, neurologists, orthopedists and neurosurgeons. To our knowledge, there was no performed MRI study on AOA cases in the literature. For this reason, especially by considering the partial-type AOA, we aimed to present additional CVJ abnormalities with MRI which might provide evidence to improve diagnostic accuracy of AOA.

Materials and Methods
A total of 11,813 cervical MRIs taken between January 2008 and October 2011, and registered in picture archiving and communication system (PACS) were scanned. AOA was detected in 40 cases. The images were acquired using 1.5 T MRI scanner (Signa Excite, GE, Milwaukee, WI, USA).

Of the 40 cases, 9 were males and 31 were females. The mean age was 40.6 years (range: 25–69 years). Ethics committee approval was obtained from Local Ethics Committee in April 2017. Written informed consent was obtained from all patients. The patients had no history of trauma or surgery, but as head-neck, back and shoulder pain, unsteadiness and dizziness.

AOA typing, anterior atlantodental interval (AADI) measurements, sagittal diameter between posterior edge of foramen magnum and atlas posterior arch (or posterior atlantodental interval, PADI) measurements were performed with sagittal FSE T1 (TR/TE: 442–460/10–16 ms, slice thickness: 2.5 mm, slice spacing: 3 mm, matrix: 512×512), FSE T2-weighted images (TR/TE: 3310–3420/82–99 ms, slice thickness: 2.5 mm, slice spacing: 3 mm, matrix: 512×512) and axial T2*-weighted gradient echo (GRE) sequence images (TR/TE: 400/14 ms, slice thickness: 2.5 mm, slice spacing: 3 mm, matrix: 512×512, flip angle: 20).

The presence of accompanying possible spine fusion anomalies, BI, tonsillar herniation and myelomalacia were noted. Suboccipital muscles [rectus capitis posterior major (RCPM), rectus capitis posterior major (RCPMA) and obliquus capitis posterior inferior (OCPI)] were evaluated. The obliquus capitis posterior superior (OCPS) muscle which is not involved in the study area was excluded from the assessment. Skull base portion and intracranial component of the VA were also evaluated. The sagittal diameter of foramen magnum above 18 mm according to the PADI measurements was considered normal.\textsuperscript{\textcircled{8,9}} AADI values greater than 3 mm were considered abnormal.\textsuperscript{\textcircled{10}} Extension of the tip of dens more than 5 mm over the Chamberlain’s line was considered as basilar invagination.\textsuperscript{\textcircled{11}} Extension of cerebellar tonsils under the opistion-basion line more than 5 mm was regarded as the tonsillar herniation.\textsuperscript{\textcircled{12}} Statistical analysis was performed using SPSS 15.0 for Windows (SPSS, Inc., Chicago, IL, USA). Data were expressed as percentages with using frequency tests.

Results
On MRI, there was a bony fusion of any part (lateral body, posterior or anterior arch) of the atlas with occipital condyle in all cases. Of the cases (n=40), bilateral fusion of the lateral bodies of atlas with occipital condyle was seen in 14 (35%) (Figure 1), complete fusion of atlas and condyle in 12 (30%) (Figure 2), symmetric fusion of lateral bodies and anterior arch with condyle in 7 (17.5%) (Figure 3), symmetric fusion of lateral bodies and posterior arch with condyle in 3 (7.5%), unilateral lateral body fusion with condyle in 3 (7.5%) and fusion of unilateral partial lateral body fusion in 1 (2.5%) cases. Atlas had hypoplastic posterior arch in 16 cases, posterior arch defect in 3 and unilateral agenesis of posterior arch in 3.

Figure 1. (a) A 48-year-old man with AOA. No bony fusion of anterior and posterior arches of the atlas with occipital condyle seen at mid-sagittal MR image, but there is severe atrophy of suboccipital muscles. (b) Parasagittal T2-weighted image shows a bony fusion of lateral body with occipital condyle (arrow).
The anomaly was accompanied with BI and C2–3 vertebrae fusion in 9 and 13 cases, respectively. PADI was < 14 mm (mean 10.46 mm) in 14 cases (35%). This value ranged from 14 to 18 mm in 20 patients and was greater than 18 mm in 6 cases. AADI was >3 mm in 10 cases (25%).

Spinal cord myelomalacia at CVJ was present in 6 patients (Figure 2). With the evaluation of suboccipital muscles, RCPM muscle was found abnormal in all cases. In 25 cases, no muscle structure was observed on MR examination and the remaining 15 cases had further findings of atrophy related to the muscle. While RCPMA muscle was symmetrically atrophic in 13 patients, 8 patients had unilateral atrophy. OCPI muscle was symmetrically atrophic in 5 patients, whereas 1 patient had unilateral atrophy (Figure 4). Tonsillar herniation was observed in 2 (5%) patients. VA abnormality was detected in 38 (95%) cases. In 30 cases, VA was in bone tunnel formed by the fusion of the atlas lateral body and occiput. VAs in 8 cases in this group showed entrapment with fused lateral bodies, odontoid

Figure 2. (a, b) A 58-year-old female presented with chronic head and neck pain. At sagittal T2-weighted images, complete form AOA is seen as fusion of anterior arch, posterior arch and lateral masses of C2 vertebra. Spinal cord myelomalacia at the C2 level and fusion of C2 and C3 vertebra also accompanied. (c) AADI increased to 5.5 mm whereas PADI decreased to 10.5 mm, revealing craniocervical instability. (d) The tip of odontoid process was projecting more than 3 mm (5.8 mm) above the Chamberline’s line, compatible with basilar invagination.

Figure 3. (a) 44-year-old woman with AOA. Fusion of the anterior arch of atlas with occipital condyle, C2–3 vertebrae fusion and suboccipital muscle atrophy were observed on sagittal T2-weighted MR image. (b) Coronal T2-weighted image shows bilateral fusion of lateral bodies (arrows) with occipital condyle. Vertebral arteries passing through a bony channel were also observed.

Figure 4. (a) A 52-year-old female with AOA. Sagittal T2-weighted MR images representing severe atrophic changes of rectus capitis posterior major (short arrow) and obliquus capitis inferior muscles (long arrow). (b) Nearly absence of rectus capitis posterior minor muscle (arrowheads). (c) Normal rectus capitis posterior major-minor (d) Obliquus capitis inferior muscles in a 52-year-old female on sagittal T2-weighted MR images.
process or brainstem formations. In 7 cases, VA was in the tunnel close to the upper surface of fused lateral body and entrapped between the occiput and cerebellum. In 20 cases, VA was extending from transverse foramen of the axis to the intracranial space by passing through the bottom of the fused lateral body. VA agenesis and advanced hypoplasia were present in 2 and 8 cases, respectively (Figure 5). While 4 cases had unilateral normal VA, bilateral normal VAs were present in only 2 cases. Additional CVJ abnormalities according to the AOA types are shown in Table 1.

Discussion

CVJ is much more mobile than any other joint of the cervical spinal column. It is necessary to know the important biomechanical properties when spinal stabilization is planned in order to ensure the proper instrumentation in the case of trauma, tumor or degenerative disease. Occiput-C1 and C1–2 joints have unique biomechanics. While bone structures are important for stabilization in occiput-C1 joint, ligamentous structures provide much more stabilization compared to the bone elements in C1–C2 joint.[13] Most of the pathological conditions affecting CVJ become symptomatic with the involvement of neural structures. Signs and symptoms are variable; typically late-onset and slowly progress, remain stable at some time and recur rarely.[14]

Although AOA is often congenital, most of the cases do not develop symptoms up to the 2nd decade of their lives. This condition is thought to be due to the gradually increase in ligamentous laxity and the increase in the degree of instability with age.[15] The incidence of AOA in our study was 0.33%; female-male ratio was 3.4. With respect to age, patients were mostly in the 4th and 5th decades.

Table 1
CVJ abnormalities according to the AOA types

<table>
<thead>
<tr>
<th>Type of AOA</th>
<th>Suboccipital muscle abnormality</th>
<th>VA abnormality</th>
<th>Increased AADI</th>
<th>Decreased PADI</th>
<th>C2–3 fusion</th>
<th>BI</th>
<th>Myelomalacia</th>
<th>Tonsillar herniation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symmetric lateral body (n=14)</td>
<td>14</td>
<td>14</td>
<td>3</td>
<td>12</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>1</td>
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<tr>
<td>Complete (n=12)</td>
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<td>12</td>
<td>6</td>
<td>12</td>
<td>7</td>
<td>6</td>
<td>4</td>
<td>-</td>
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<td>5</td>
<td>1</td>
<td>5</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Symmetric, posterior arch and lateral body (n=3)</td>
<td>3</td>
<td>3</td>
<td>-</td>
<td>2</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
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<td>Unilateral, lateral body (n=3)</td>
<td>3</td>
<td>3</td>
<td>-</td>
<td>2</td>
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<td>-</td>
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<tr>
<td>Unilateral, partial lateral body (n=1)</td>
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<td>1</td>
<td>-</td>
<td>1</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Total (n=40)</td>
<td>40 (100%)</td>
<td>38 (95%)</td>
<td>10 (25%)</td>
<td>34 (85%)</td>
<td>13 (33%)</td>
<td>9</td>
<td>6 (15%)</td>
<td>2 (5%)</td>
</tr>
</tbody>
</table>

Menezes[5] reported that anterior arch assimilation with BI is accompanied with instability explained by the restricted movement caused by weakening of supportive myoligamentous structure resulting from transfer of craniospinal first mobile segment into the C1–2 vertebrae, atlantoaxial joint weakness and progressive atlantoaxial subluxation. According to Wackenheim, cranial translocation of odontoid process is the main criterion for AOA, and other skeletal anomalies are accompanied, especially C2–3 vertebrae fusion.[16]

Gholve et al.[17] identified 2 types of BI directly causing brainstem compression and showing association with Chiari malformation and reduction in posterior cranial fossa and showed that the risk of atlantoaxial instability is high in cases with fused C2–3 vertebrae. In our study group, AOA was accompanied by posterior arch hypoplasia (n=16), midline defect of posterior arch (n=3) and unilateral agenesis of the posterior arch (n=3). BI was present in 9 (22.5%) cases and C2–3 vertebrae fusion in 13 (32.5%) cases. Both anomalies were more common in complete form AOA cases. Isolated anterior arch fusion was not observed in the study group. Of the cases in which BI was detected, 6 had complete form fusion, 2 had symmetric lateral body fusion, and one showed symmetric fusion of both anterior arch and lateral body. In other words, 7 of 9 cases with BI had anterior arch involvement. 4 of the cases accompanied by BI had C2–3 vertebrae fusion. Tonsillar herniation was detected in 2 cases which had isolated symmetric lateral body and lateral body with anterior arch type fusion accompanied with hypoplasia of the posterior arch.

AOA may narrow foramen magnum diameter and lead to neurological complications resulting from spinal cord compression.[18,19] BI and dorsal displacement of odontoid process typically results in reduced anteroposterior diameter of foramen magnum and significant compression on craniomedullar junction (CMJ). Sudden death cases associated with AOA have been reported in the literature.[19] Although increased AADI indirectly supports the possibility of instability, this increase may remain constant or there may not be neural compression. Therefore, increased AADI does not indicate neurological abnormality. On the other hand, decrease in PADI does not mean a neurological abnormality, either. However, the decrement of PADI can cause anterior spinal, VA and basilar artery failure without leading to direct spinal cord compression.[20] It has been reported that PADI value is correlated with the presence of paralysis and its severity. It is the most important potential indicator of neurological recovery after surgery in patients with rheumatoid arthritis.[21]

Greenbery[20] reported that PADI value of 14 mm or less means spinal cord compression; 14–18 mm means possible spinal cord compression, and 18 mm or above means no compression. In our study group, PADI was below 14 mm in 14 cases and mostly associated with complete form AOA. In 20 cases, PADI was measured between 14 to 18 mm. AADI was greater than 3 mm in 10 cases and mostly (6 cases) were associated with complete form AOA. 7 cases with decreased PADI (7/14) also had increased AADI. In 5 of 6 cases with myelomalacia at the CMJ, PADI was lower than 14 mm, and in 3 cases AADI was above 3 mm. In 7 cases (7/14) with C2–3 vertebrae fusion, PADI was below 14 mm, in 5 cases between 14–18 mm and in 1 case PADI was above 18 mm. In all cases, in which BI was accompanied with myelomalacia, PADI was lower than 14 mm and, in this group C2–3 vertebrae fusion was present in 2 cases.

Wackenheim[14] stated that assimilation was always accompanied with VA abnormalities. Wang et al.[22] reported 4 different types of VA at the CVJ level in AOA. In Type 1, VA extends into the spinal canal and foramen magnum at the bottom of the C1 posterior arch immediately after leaving transverse foramen of the axis. VA course is below the assimilated C1 lateral mass. In Type 2, VA enters into the spinal canal and foramen magnum below the assimilated C1 posterior arch. In Type 3, VA passes through transverse foramen of the atlas, enters into osseous foramen generated by fused atlas and occiput. In Type 4, VA is unilaterally agenetic. Tubbs et al.[23] examined VAs in the skulls of 5 adult cadavers with AOA and reported the presence of abnormal osseous pathway into the cranium that mostly had posterior arch or hemiarch fusion. In this study, VA was abnormal in 38 cases (95%), VAs passing through the tunnel by crossing the bone bar formed by the fused occiput and lateral mass of atlas after leaving the transverse foramen of the axis was present in 30 cases and they showed Type 3 feature according to Wang’s classification. In 20 cases, VAs were featured as Type 1 and 2. Agenetic VA (Type 4) and advanced hypoplastic VA was detected in 2 and 8 cases respectively. In 8 cases showing Type 3 VA property (in 6 cases VA was accompanied with BI), VAs showed entrapment between odontoid process, fused lateral mass and the brain stem. Again, VAs of 7 cases in this group were in the tunnel located near the upper surface of fused lateral mass and entrapped between the occiput and cerebellum. VA was completely normal in 2 cases and both cases had fusion of anterior arch and lateral mass accompanied by posterior arch hypoplasia. While right and left VA exhibited

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the same type of abnormality in most of the cases, in some cases, different types of VA abnormality were present in the same patient.

As well as the reduction of the luminal diameter caused by lateral mass protrusion to the foramen magnum, compression of the first cervical nerve affect suboccipital muscles and lead to abnormal head position and unstable walking. Bodon et al. examined the CVJ and suboccipital muscle changes in cadaver skull with occipitalization. In some studies, in cases with chronic head and neck pain, dizziness and imbalance complaints, atrophic changes were identified with MRI in suboccipital muscles especially in RCPM and RCPMA muscles. In our study, RCPM, 1 of the suboccipital muscles, was abnormal in all patients. There was no identifiable muscular tissue in 20 cases. In 15 cases, RCP was severely atrophic. This condition may arise as a result of compression of the first cervical nerve and is also attributed to the atrophy of the muscle that is effective in flexion and extension of the head resulting from the movement restriction caused by assimilation.

With review of the study results, we realized that as the severity and distribution of bony fusion in AOA increased, the frequency of accompanying CVJ abnormalities also increased. In complete and symmetric lateral body/arch fusions, various CVJ abnormalities were accompanied, while partial and/or unilateral forms had less abnormalities. We also noticed that the type and severity of the fused bony segment can predict the nature of CVJ abnormality. In this context, cases with VA abnormality mostly had lateral body involvement. CVJ abnormality for suboccipital muscles was remarkably seen in all cases whatever the type and severity of assimilation.

Our study has several limitations. First, because the cervical MRIs were obtained by standard protocols, OCPS muscle, not included in the study area, could not be evaluated. Second, since axial plan images did not include the CVJ level in all cases, vertebral artery was only evaluated on sagittal images in some patients. To include the CVJ level in all cases, vertebral artery was scrutinized in 20 cases. In 15 cases, RCP was severely atrophic. This condition may arise as a result of compression of the first cervical nerve and is also attributed to the atrophy of the muscle that is effective in flexion and extension of the head resulting from the movement restriction caused by assimilation.

Conclusion
Considering AOA has various types, ranging from complete to a minute bony fusion and according to the fused bony part of the atlas, cases with AOA can be simply overlooked. In order to improve the diagnostic rate of AOA and thus minimize the unnecessary radiologic examinations and reduce the complications especially in the patients scheduled for surgery, it is useful and important for the radiologists, neurologists, orthopedic surgeons and neurosurgeons to display severe atrophy of suboccipital muscles, VA abnormality, BI, C2–3 vertebral fusion, reduction of foramen magnum sagittal diameter, and myelomalacia.

References


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