Imaging findings for Atlanto-occipital assimilation with multiple cervical vertebral anomalies in a Beagle dog: A 2 year follow-up

Jaehwan Kim, Misuk Chae, Kidong Eom*

Department of Veterinary Medical Imaging, College of Veterinary Medicine, Konkuk University, Neungdong-ro, Gwangjin-gu, Seoul, Republic of Korea

*Corresponding author: eomkd@konkuk.ac.kr


Abstract: A 3-year-old male Beagle dog was presented for a physical examination, which revealed a mild stiff neck that was affecting movement. The imaging modalities led to our diagnosis of an asymmetric fusion of the occipital condyle and atlas wing consistent with Atlanto-occipital assimilation (AOA). An incomplete ossification of the atlas and axis, dysplastic dens, and a blocked vertebra were also noted. During a two-year follow-up, the dog showed no other clinical signs or disease progression. To our knowledge, this is the first imaging description of AOA in veterinary medicine. We recommend a careful CT (computed tomography) and an MRI (magnetic resonance imaging) evaluation in cases of neck pain and cervical myelopathy, and inclusion of AOA among the differential diagnoses, with the awareness that its clinical signs resemble those of other canine diseases.

Keywords: Atlanto-occipital assimilation; Atlanto-occipital fusion; atlas occipitalisation; cervical vertebral anomaly; dog

Atlanto-occipital assimilation (AOA) is defined as the fusion of the atlas with the occipital bones, and is variously referred to as Atlanto-occipital fusion, occipitocervical synostosis, atlas occipitalisation, or cranio cervical fusion (Al-Motabagani et al. 2006). It is a rare skeletal variation classified as a type of transitional vertebra of the cranio cervical junction (CCJ) (Ciolkowski et al. 2014). The CCJ is clinically important in small and toy-breed dogs, because diseases affecting this region may cause clinical signs, as in a Chiari-like malformation (Dewey et al. 2013), a bipartite atlas (Wrzosek et al. 2014), dens dysplasia (Zaki 1980), and Atlanto-occipital overlapping (Cerda-Gonzalez et al. 2009). Although it is one of the most common cranio cervical abnormalities reported in human medicine (Kim et al. 2013), the clinical importance of AOA and its imaging characteristics have not been reported in veterinary medicine. The purposes of this case report are to describe the imaging features of AOA and to investigate its clinical significance in dogs.

Case description

A 3-year-old male Beagle dog was presented for a physical check-up. The dog did not show any clinical signs, nor did it have any previously diagnosed disease. On physical examination, the dog resisted ventral neck flexion. A neurological examination and blood work revealed no significant findings. We took cervical vertebral radiographs to screen for a possible abnormality in the cervical vertebrae, using a commercially available X-ray machine (Titan 2000M; COMED Medical Systems,
Seoul, Republic of Korea) (Figure 1). The right lateral and ventrodorsal projection radiographs were taken under 72 kVp, 200 mA, 0.035 sec with the dog in a conscious state. On the lateral view, anomalies of the occipital bone and the first three cervical vertebrae were present. The summation effect of the wings of the atlas was not identifiable (Figure 1A). The spinocephalic process of the axis was not fused with the lamina, and the vertebral body was separated into two parts by incomplete ossification. An intervertebral disc-space-like structure was visible in the middle of the axis between the unfused segments. A ventral bony spur was visible between the axis and the 3rd cervical vertebra, consistent with a blocked vertebra resulting from the fusion of the adjacent vertebral bodies. On the ventrodorsal projection, the bilateral wings of the atlas were asymmetric, and the left wing was abnormally fused with the ipsilateral occipital condyle (Figure 1B). A remarkable radiolucent space observed at the neural arch of the atlas was considered to represent the incomplete ossification of the atlas. The left transverse process of the atlas was abnormally prolonged compared to that on the contralateral side. Computed tomography (CT) and magnetic resonance imaging (MRI) were performed for further evaluation of the CCJ and adjacent cervical spinal cord.

For the CT scan, anaesthesia was induced with propofol (6 mg/kg, i.v. Provive 1%; Myungmoon Pharmaceutical Co., Seoul, Republic of Korea) and maintained by endotracheal intubation with 1.5% isoflurane (Foran solution; Choongwae Pharma Corporation, Seoul, Republic of Korea) in 100% oxygen. The CT scan (Brivo CT385; GE healthcare system, Waukesha, USA) was performed with the dog in ventral recumbency using a pre-contrast bone algorithm, without gantry tilting, and with the following specifications: 100 kVp, 200 mAs, and 1.25 mm slice thickness. On the transverse images, the left wing of the atlas was fused with the ipsilateral occipital condyle (Figure 2A). At the level of the atlantoaxial junction, a blunted, dysplastic odontoid process with an incomplete ossification of the atlas resulted in the narrowing of the vertebral canal (Figure 2B). The atlas lacked ossification at the dorsal suture of the neural arch, and at the bilateral sutures of the intercentrum. On the sagittal plane, the tip of the odontoid process was identified as blunted and dysplastic without compression of the adjacent spinal cord. The shape of the occipital bone and the size of foramen mag-

Figure 1. Lateral (A) and ventrodorsal (B) cervical vertebral radiographs. The anomalous and misaligned cranio-cervical junction is visible. Summation of the wings of the atlas and odontoid process is not visible on the lateral view. The axis shows the incomplete ossification of the vertebral body (white arrowhead) and the spinocephalic process (black arrow). A ventral bony spur between the axis and the 3rd cervical vertebra, consistent with a blocked vertebra, is identified (open white arrowhead). On the ventrodorsal projection, the atlas is bipartite, and the left wing (black asterisk) is fused with the ipsilateral side of the occipital condyle (white arrow). A radiolucent space is present at the level of the atlas that consistent with the incomplete ossification of the neural arch of the atlas (black arrowheads). Additionally, a prolonged right transverse process of the axis (white asterisk) is noted.
num were normal. The remaining findings were confirmed to be similar to those in the radiographs. The MRI was performed using a 1.5-Tesla system (Signa HDxt; GE Healthcare System, Waukesha, USA) consecutively after the CT scan. The dog was positioned in dorsal recumbency on the 8-channel phased-array spine coil. The parameters of the MRI scans for the T2-weighted transverse images were as follows: relaxation time, 3 800 msec; echo time, 11.6 msec; flip angle, 160°; slice thickness, 3 mm; and interslice gap, 3.3 mm, and for the T2-weighted sagittal images, they were as follows: relaxation time, 3 500 msec; echo time, 105.3 msec; flip angle, 160°; slice thickness, 3 mm; and interslice gap, 3.3 mm. Our examination did not reveal any distinct spinal cord damage, cervical syringohydromyelia, or compression associated with AOA and spinal canal stenosis. We identified crowding of the spinal cord by the atlas and odontoid process, and
the Atlanto-occipital malalignment (Figure 3). On the sagittal plane, a disc-like structure was located at the level of the segmented body of the axis, and a mild protrusion of the intervertebral disc was visible between the axis and the 3rd cervical vertebra. The herniated and degenerative disc compressed the adjacent epidural fat, without any distinct spinal cord compression. A dynamic MRI with the dog in a flexed neck position did not reveal any spinal cord compression or instability at the craniocervical junction (Figure 3D).

Follow-up

Two years later, follow-up radiographs and an MRI were performed at the owner’s request (Figure 4). A dynamic study of the radiographs depicting the dog with a stressed flexion of the head in an extended neck position did not reveal evidence of the remarkable instability associated with AOA in humans. There were no visible degenerative changes such as the presence of osteophytes and enthesophytes. A follow-up MRI scan was

Figure 3. Magnetic resonance imaging findings at the level of the craniocervical junction. (A) The left occipital condyle (asterisk) and wing of the atlas are fused (arrow). (B) At the level of the Atlanto-axial junction, the vertebral canal is narrowed due to the abnormalities of atlas and axis, but compression of the spinal cord (sc) is not identified. (C) On the sagittal plane, a disc-like structure is identified between the abnormally segmented vertebral body of the axis (arrowhead). The axis and the 3rd cervical vertebra are fused ventrally, causing the narrowing of the disc space (open arrowhead). (D) A dynamic MRI study with the dog in a flexed cervical position did not reveal any spinal cord compression or instability at the craniocervical junction.

(A) T2-weighted, transverse plane at the level of the Atlanto-occipital junction; (B) T2-weighted, transverse plane at the level of the Atlanto-axial junction; (C) T2-weighted, mid-sagittal plane; (D) T2-weighted, mid-sagittal plane, flexion stressed view.
performed using the same conditions as the previous one, and no remarkable progression or change was observed. The signals indicating the presence of an intervertebral disc on the cervical vertebra were similar to those of the previous MRI scan. As a result, the dog was diagnosed with a subclinical, non-progressive AOA with odontoid process dysplasia and a blocked vertebra concurrent with the incomplete ossification of the atlas and the axis. The dog was discharged without administration of any special treatment or a prescription.

DISCUSSION AND CONCLUSION

AOA is one of the most common disorders of the CCJ and can be both congenital in origin or acquired during the lifetime, with an incidence ranging from 0.1% to 2.7% in humans (Sharma et al. 2017). Most cases are congenital in origin, caused by fusion of the cranial part of the first sclerotome segment with the caudal part of the fourth occipital sclerotome segment (Kim et al. 2013). In addition, unlike other vertebrae, the atlas has three ossification centres which can result in a variety of congenital disorders in the CCJ region (Wrzosek et al. 2014). Most congenital conditions affecting the CCJ in dogs are characterised by complex malformations that affect more than a single site. Rarely, this condition is acquired and involves the formation of a fibrous band, caused by infection or trauma, resulting in the fusion of the occipital condyle with the atlas. Anomalies of the cervical vertebra often occur multifocally and are accompanied by malfor-
mations of other spinal elements. The present case is considered congenital because it was accompanied by multiple cervical vertebral malformations and the fusion was caused by the bony tissue rather than a fibrous band. The multimodal imaging did not identify any abnormal findings in the adjacent tissues. The lack of clinical symptoms or progression also supported our conclusion that this condition was congenital.

AOA is classified into two subcategories: complete and partial types. Complete AOA involves the fusion of the entire atlas, whereas partial AOA involves an asymmetric fusion of part of the atlas to the occipital bone. The complete type may cause various clinical symptoms because it induces more severe stiffness. In the present case, as only the left side of the atlas fused with the occipital condyle, consistent with partial AOA, it is probably a subclinical asymptomatic condition that had been present for a long time. On follow-up, the vertebral instability or disc degeneration was barely detected, implying that a relatively stable joint had formed despite several anomalies concurrent with the AOA. It is also possible that the limited cervical movement associated with the blocked vertebra and AOA may have delayed any progression of the disease. The follow-up confirmed that despite various craniocervical malformations, the dog’s condition was subclinical. Considering that AOA is the most commonly occurring anomaly of the CCJ in humans (Al-Motabagani and Surendra 2006; Ciolkowski et al. 2014; Nimje and Wankhede 2014), it is possible that a large percentage of dogs with AOA are subclinically affected and show no clinical signs throughout their lifetime.

Generally, a malformation of the CCJ is of considerable consequence due to its proximity to the spinal cord and medulla oblongata, which increases the risk of neurological compression. AOA induces various clinical symptoms, the severity of which ranges from a subclinical form to headaches, neck pain, weakness, ataxia, torticollis, and tetraplegia. The neurologic signs are mostly induced by the narrowing of the foramen magnum and vertebral canal that may compress the spinal cord (Kim et al. 2013). Fusion of the CCJ results in absolute or partial immobility and usually leads to the compensatory hypermobility of the adjacent joints, especially the atlantoaxial joints (Al-Motabagani and Surendra 2006). It has been previously reported that approximately 50% of human patients with AOA develop atlantoaxial instability and concurrent subluxation (Sharma et al. 2017). Since atlantoaxial instability is a relatively common cervical anomaly in dogs, careful attention should be paid to detect the presence of the concurrent AOA. In the present case, there was no evidence of an atlantoaxial instability or an adjacent cervical vertebral subluxation seen on both the dynamic cervical radiographs and MRI images with the dog in a flexed neck position. Although we expected the condition to be progressive because we had confirmed the presence of a blocked vertebra, disc degeneration, and a mild protrusion, the serial MRI scans did not show any remarkable evidence of progression. This implies that the presence of a subluxation and instability detected via a dynamic study may be helpful for predicting the potential severity and likelihood of the progression of the disease.

AOA is a part of an occipitoatlantoaxial malformation associated with many conditions, such as atlantoaxial instability, odontoid process dysplasia, incomplete ossification of the atlas, a blocked vertebra, cervical ribs, urinary tract anomalies, spina bifida, and a cleft palate in humans (Wang et al. 2009; Ciolkowski et al. 2014). In the present case, dens dysplasia, a blocked vertebra, and the incomplete ossification of the atlas and axis were confirmed to be concurrent with AOA, as in human medicine. Odontoid process dysplasia is a major cause of atlantoaxial instability, and is clinically significant in dogs due to the high incidence of the concurrent abnormal dorsal transverse ligament (Warren-Smith et al. 2009). Ossification of the atlas is completed at approximately 4 months of age. The incomplete ossification of the atlas occurs in approximately 10% of dogs and is a predisposing factor in the development of atlantoaxial instability (Parry et al. 2010; Warren-Smith et al. 2009). However, our patient’s condition was confirmed to be subclinical despite the odontoid process dysplasia accompanied by the incomplete ossification of the atlas. Nevertheless, for suspected AOA in dogs, whether clinically significant or not, we recommend whole-body screening to detect other congenital anomalies because most congenital conditions affecting the CCJ are accompanied by a combination of complex malformations rather than the sole occurrence of AOA.

In conclusion, we believe this to be the first imaging description of AOA in veterinary medicine. AOA is a type of transitional vertebra, characterised
by the fusion of the occipital bone and the atlas. Because AOA potentially occurs with various cervical vertebral anomalies, we recommend a careful evaluation using CT and MRI. Moreover, in cases of neck pain and cervical myelopathy, AOA should be considered among the differential diagnoses, because it might induce clinical signs similar to those of other previously-reported canine cervical myelopathy.

Conflict of interest

The authors declare no conflict of interest.

REFERENCES


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