Open Veterinary Journal, (2017), Vol. 7(1): 65-69
ISSN: 2226-4485 (Print)
ISSN: 2218-6050 (Online)
Case Report
DOI: http://dx.doi.org/10.4314/ovj.v7i1.10

Submitted: 08/10/2016 Accepted: 20/03/2017 Published: 25/03/2017

Craniocervical junction abnormalities with atlantoaxial subluxation caused by ventral subluxation of C2 in a dog

Harumichi Itoh[1], Kazuhito Itamoto[1,*], Shotaro Eto[2], Tomoya Haraguchi[1], Shimpei Nishikawa[1], Kenji Tani[2], Yoshiki Itoh[3], Masato Hiyama[2], Toshie Iseri[3], Munekazu Nakaichi[3] and Yasuho Taura[2]

[1]*Department of Small Animal Clinical Science, Joint Faculty of Veterinary Medicine, Yamaguchi University, 1677-1 Yoshida, Yamaguchi City, Yamaguchi, 753-8511, Japan*
[2]*Department of Veterinary Surgery, Joint Faculty of Veterinary Medicine, Yamaguchi University, 1677-1 Yoshida, Yamaguchi City, Yamaguchi, 753-8511, Japan*
[3]*Laboratory of Veterinary Radiology Yamaguchi University, 1677-1 Yoshida, Yamaguchi City, Yamaguchi, 753-8511, Japan*

Abstract
Craniocervical junction abnormalities with atlantoaxial subluxation caused by ventral subluxation of C2 were diagnosed in a 6-month-old female Pomeranian with tetraplegia as a clinical sign. Lateral survey radiography of the neck with flexion revealed atlantoaxial subluxation with ventral subluxation of C2. Computed tomography revealed absence of dens and atlanto-occipital overlapping. Magnetic resonance imaging showed compression of the spinal cord and indentation of caudal cerebellum. The diagnosis was Chiari-like malformation, atlantoaxial subluxation with ventral displacement of C2, atlanto-occipital overlapping, and syringomyelia. The dog underwent foramen magnum decompression, dorsal laminectomy of C1, and ventral fixation of the atlantoaxial joint. Soon after the operation, voluntary movements of the legs were recovered. Finally, the dog could stand and walk without assistance. The dog had complicated malformations at the craniocervical junction but foramen magnum decompression and dorsal laminectomy for Chiari-like malformation, and ventral fixation for atlantoaxial subluxation resulted in an excellent clinical outcome.
Keywords: Atlantoaxial subluxation, Atlanto-occipital overlapping, Chiari-like malformation, Craniocervical junction abnormalities, Foramen magnum decompression.

Introduction

Atlantoaxial subluxation (AAS) is a disorder of C1-C2 causing impairment of stability; the causes of instability are associated with aplasia, hypoplasia, dorsal angulation, or non-union of the dens with the C2, and congenital absence of the transverse ligament (Thomas *et al*., 1991). Recently, craniocervical junction abnormalities (CJA) were identified as complicated congenital malformations at the region of the caudal occiput and first two cervical vertebrae. Chiari-like malformation (CLM) appears commonly in CJA patients, and other abnormalities are atlanto-occipital overlap (AOO), dorsal constriction at C1/C2, and AAS (Dewey *et al*., 2013).

Most cases of AAS subluxate C2 dorsally and many surgical approaches for AAS with dorsal subluxation have been reported. It have been suggested ventral stabilization may be safer than dorsal stabilization of the atlas and axis (Aikawa *et al*., 2013) and the application of ventral pins and polymethylmethacrylate has been used successfully in the surgical treatment of congenital and traumatic AAS with dorsal subluxation of C2 (Schulz *et al*., 1997). However, there has been no report of AAS with ventral subluxation of C2 and surgical treatment in dogs. The following case report describes AAS with ventral subluxation of C2.

Case Details

A 6-month-old female Pomeranian presented (body weight = 1.0 kg) with tetraplegia for 3 days and was referred to the Animal Medical Centre of Yamaguchi University. The neurological examination showed loss of proprioception and upper motor neuron paresis of fore- and hindlimbs. Lateral radiography of the cervical region revealed ventral subluxation of C2. Ventrodorsal radiography showed a deficit of the dens at C2. Stress radiography of the cervical region with ventral flexion revealed that the distance between the caudal margin of dorsal arch of C1 and the cranial margin of spinal process of C2 was 3.7 mm (Fig. 1) (normal range <4 mm). Sagittal MRI of the cranial cervical spine revealed severe compression of the spinal cord at C1 to C2 level and indentation of the caudal cerebellum (Fig. 2). Sagittal computed tomography (CT) revealed an occipital bony defect, and the cranial-most aspect of the dorsal arch of C1 was inserted into the intracranial region from the foramen magnum (Fig. 3).

***Corresponding Author:** Kazuhito Itamoto. Yamaguchi University, Animal Medical Center 1677-1, Yoshida, Yamaguchi-shi, Japan. Email: kaz2356@yamaguchi-u.ac.jp

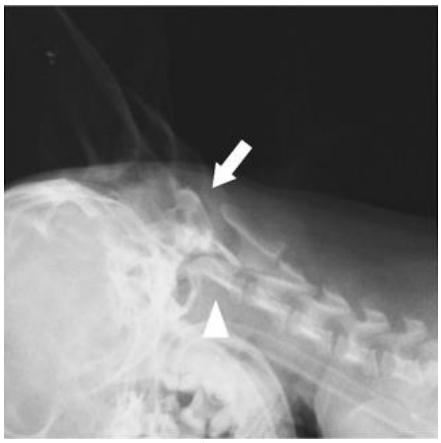

Fig. 1. Stress radiography of the cervical region with ventral flexion. The space between the dorsal lamina of the C1 and the dorsal spinous process of the C2 was 3.7mm (arrow). Ventral subluxation of C2 (arrowhead).

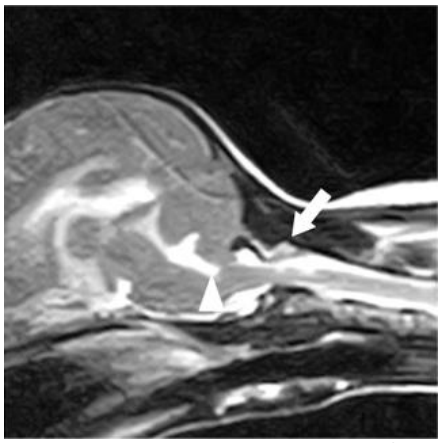

Fig. 2. Sagittal MRI image (T2-weighted) of the cranial cervical spine. Severe compression of spinal cord at C1 to C2 level (arrow) and indentation of caudal cerebellum (arrowhead).

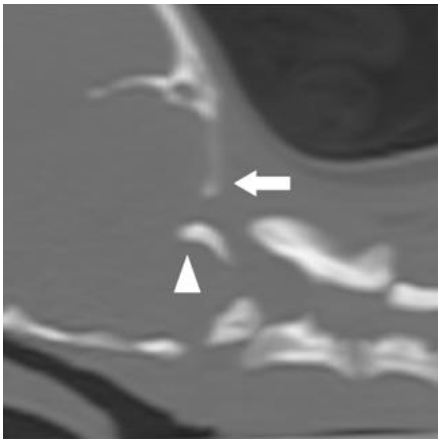

Fig. 3. Sagittal CT image of the cervical region. Occipital bony defect (arrow). Cranial-most aspect of dorsal arch of the C1 was inserted to intracranial region from foramen magnum (arrowhead).

According to these examinations, the patient was diagnosed as AAS complicated by AOO, CLM, and syringomyelia. The operative approach was divided into two phases. First, in order to treat AOO, CLM and syringomyelia, foramen magnum decompression (FMD) and dorsal laminectomy of C1 were planned to decompress the cerebellum and spinal cord. Then, in order to stabilize the atlantoaxial joint, ventral fixation of the joint was planned for the second phase. The first phase of surgery (FMD and dorsal laminectomy of C1) was performed on the 7th day after the first consultation. Anaesthesia was induced with propofol IV (7 mg kg^{-1} propofol; Intervet, Tokyo, Japan). An endotracheal tube was inserted and isoflurane (Isoflu; DS Pharma Animal Health Co, Osaka, Japan) anaesthesia was maintained at 1-2% using a low-flow, semi-closed circuit. Analgesia was performed by preoperative intara muscular administration of ketamine hydrochloride (5 mg kg^{-5} Ketalar; Daiichi Sankyo, Tokyo, Japan) and bolus administration of fentanyl (Fentanyl; Daiichi Sankyo, Tokyo, Japan). In addition, continuous rate infusion of fentanyl citrate (5 to 30 µg/kg/min) was used for intra-operative analgesia.

The patient was positioned in sternal recumbency with the neck ventroflexed. A dorsal midline incision was made extending from external occipital protuberance cranially to the second cervical vertebra caudally. The superficial dorsal cervical musculature was separated at the median raphe, exposing the underlying paired biventer cervicis muscles. The paired muscles were separated at the midline, exposing the rectus capitis dorsalis muscles. The caudal aspects of the rectus capitis dorsalis muscles were removed from the cranial half of the second cervical vertebra and the muscle bellies were split at the midline. The cranial aspects of the rectus capitis dorsalis muscles were then sharply incised from the nuchal crest, exposing the caudal portion of the occiput and the arch of the atlas.

The occiput and the dorsal aspect of the C1 vertebra were removed using a high-speed air drill with round drill. The dorsal atlanto-occipital membrane was incised and expansile duroplasty was performed by using artificial dura mater (Gore-Tex; Japan Gore, Tokyo, Japan) (Fig. 4). Closure was routine. Postoperative analgesia was performed appropriately by intramusucular administration of morphine hydrochloride (0.5 mg kg^{-1} Morphine; Takeda, Osaka, Japan). Ventral fixation of the atlantoaxial joint was performed to stabilize it on the 14th day after the first consultation. Anaesthesia and analgesia were performed as in the previous surgery. The dog was placed in dorsal recumbency with the neck hyperextended. A ventral midline skin incision was made over the cranial half of the neck.

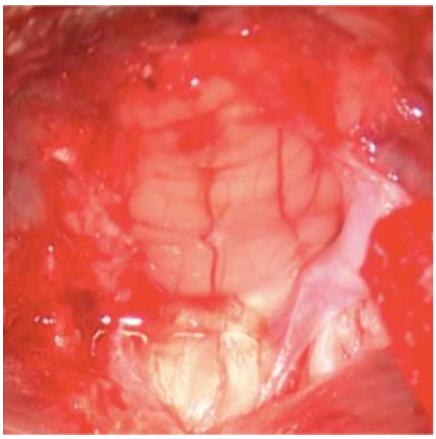

Fig. 4. The dorsal atlanto-occipital membrane were incised and expansile duroplasty by using artificial dura mater.

The sternohyoideus muscles were divided in the midline and the trachea and oesophagus were retracted toward the dog's left side. The larynx was also retracted toward the left, once the attachments of the right sternohyoideus muscle were severed close to the larynx. The thyroid gland, recurrent laryngeal nerve, right vagus, and carotid were identified and protected throughout the surgery. The longus colli muscle was elevated from the ventral surface of the axis and atlas and retracted.

Two threaded-pins (0.9 × 75 mm; IMEX, Tokyo, Japan) were placed across each of the ventral articular facets between the axis and atlas. Then, removal of cartilage of the atlantoaxial joint and autografting of cancellous bone collected from the humerus were performed to create bone union at the atlantoaxial joint. Finally, implanted pins were fixed by using bone cement (Osteobond; Zimmer, Tokyo, Japan) (Fig. 5).

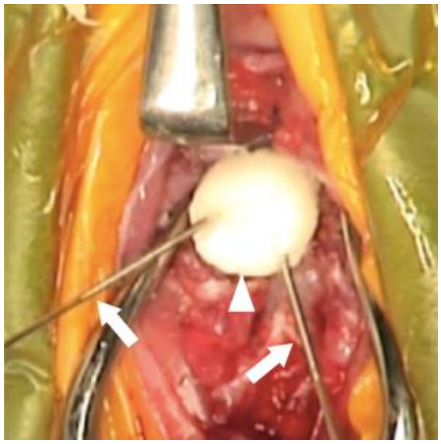

Fig. 5. Two threaded-pins are placed across each of the ventral articular facets between the axis and atlas (arrows). Implanted pins are fixed by using bone cement (arrowhead).

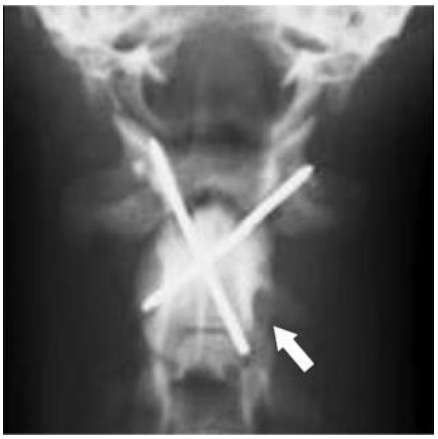

Fig. 6. 134 days after the first consultation, ventro-dorsal radiograph of the cervical region revealed no loosening of thread pins and bone cement (arrow).

The dog stood and walked by herself and left the medical centre. Ventro-dorsal radiograph of the cervical region in 134 days after the first consultation revealed no loosening of thread pins and bone cement (Fig. 6).

Discussion

In AAS, dorsal subluxation of the C2 into the vertebral canal causes direct compressive and concussive effects on the cervical spinal cord. Usually, the subluxation of C2 would occur dorsally in accordance with the direction of cervical flexion when stability is lost. However, in this case, C2 subluxated ventrally. We found no previous report about AAS with ventral subluxation of C2 in dogs.

In human cases, a form of AAS similar to this case is known as rotatory subluxation type IV (rotatory subluxation with posterior displacement of the atlas). Rotatory subluxation type IV is rare, but secondary rotatory subluxation type IV has been reported in patients with rheumatoid arthritis (Kauppi, 1994). If the erosions of the dens increase progressively in rheumatoid arthritis patients, the dens may shorten, disappear, or fracture. If the dens does not limit the posterior movement of the atlas during extension, the anterior atlas arch may glide over the whole anterior part of the axis. However, not of all AAS patients with rheumatoid arthritis and erosion of the dens shows rotatory subluxation type IV and details of mechanism about rotatory subluxation type IV have not been investigated even now.

On the other hand, we found no previous report of AAS with ventral subluxation of C2 even in dogs with rheumatoid arthritis. Anatomically, atlantoaxial joint ligaments in dogs constructed by four major ligaments. Apical, left and right alar, transverse and dorsal atlantoaxial ligaments are known commonly. A previous report concluded alar ligaments are the most

important in prevending dorsal angulation (Reber *et al*., 2013).

According to these issues, it is suggested that absence of dens allows dorsal angulation of axis by lose their functional ability of alar ligaments. In fact, some AAS patients with dorsal subluxation of C2 with deficit of the dens have been reported (Patton *et al*., 2010; Stigen *et al*., 2013). Therefore, the patient who has AAS with absence of dens shows higher opportunity to dorsal subluxation of C2 than normal AAS patients. Compared to these previous reports, the patient in this case report shows ventral subluxation. The exact details why this case report showed ventral subluxation are unclear. However, further investigation about this issues and follow-up to the case is necessary.

In this case, AOO was also observed. AOO can be seen in toy and small breed dogs (Cerda-Gonzalez and Dewey, 2010). A previous report described 4 cases of AOO. In this report, 3 cases had AAS (Cerda-Gonzalez *et al*., 2009). A case report of surgical stabilization for AOO was reported. The case was performed with combined FMD with cranioplasty and stabilization of the atlanto-occipital junction (Dewey *et al*., 2009). We also performed FMD and dorsal laminectomy of C1.

The treatment of AAS has long been performed and many treatment methods have been reported. Surgical treatment with dorsal and ventral technique has been reported. These include the dorsal Kishigami AATB technique (Pujol *et al*., 2010), dorsal cross-pinning (Jeffery, 1996), and others. Ventral surgical treatment using transarticular lag screws/pins (Riedinger *et al*., 2015) and screws, and polymethylmethacrylate (Platt *et al*., 2004; Schulz *et al*., 1997), have better clinical outcomes than dorsal surgical treatments (Aikawa *et al*., 2013). In this case, we chose ventral fixation to treat AAS. Usually, dorsal fixation of the atlantoaxial joint needs traction on C1 in a dorsal direction when there is dorsal subluxation of C2. However, in this case, AAS was difficult to correct because of the opposite direction of subluxation. In addition, we planned FMD and dorsal laminectomy at the first operation.

In this case, AAS was concomitant with AOO, CLM, and syringomyelia. The deficit of dens, foramen magnum hypoplasia, and indentation of the caudal cerebellum were also observed. The occipital region of the skull and the first two cervical vertebrae develop together embryologically (Dewey *et al*., 2013). Therefore, atlantoaxial instability sometimes has other concurrent abnormalities. For example, occipital dysplasia and AOO lead to CLM. Malformations of the dens and absence of the transverse ligament also lead to atlantoaxial joint instability. These malformations occur together. Recently, these complicated malformations were named “Craniocervical Junction Abnormalities” (Cerda-Gonzalez and Dewey, 2010). Our case also had combined AAS, CLM, and AOO. Therefore, it was categorized as a CJA.

In conclusion, this was the first reported case of AAS with ventral subluxation and complicated malformations of CJA. The combination of FMD and ventral fixation of the atlanto-axial joint resulted in an excellent clinical outcome.

References

Aikawa, T., Shibata, M. and Fujita, H. 2013. Modified ventral stabilization using positively threaded profile pins and polymethylmethacrylate for atlantoaxial instability in 49 dogs. Vet. Surg. 42, 683-692.

Cerda-Gonzalez, S. and Dewey, C.W. 2010. Congenital diseases of the craniocervical junction in the dog. Vet. Clin. North Am. Small Anim.Pract. 40, 121-141.

Cerda-Gonzalez, S., Dewey, C.W., Scrivani, P.V. and Kline, K.L. 2009. Imaging features of atlanto-occipital overlapping in dogs. Vet. Radiol. Ultrasound 50, 264-268.

Dewey, C.W., Cerda-Gonzalez, S. and Scrivani, P.V. 2009. Case report --surgical stabilization of a craniocervical junction abnormality with atlanto-occipital overlapping in a dog. Compend. Contin. Educ. Vet. 31, E1-6.

Dewey, C.W., Marino, D.J. and Loughin, C.A. 2013. Craniocervical junction abnormalities in dogs. N. Z. Vet. J. 61, 202-211.

Jeffery, N.D. 1996. Dorsal cross pinning of the atlantoaxial joint: new surgical technique for atlantoaxial subluxation. J. Small. Anim. Pract. 37, 26-29.

Kauppi, M. 1994. A method for classification of the posterior atlanto-axial subluxation. Clin. Rheumatol. 13, 492-495.

Patton, K.M., Almes, K.M. and de Lahunta, A. 2010. Absence of the dens in a 9.5-year-old rottweiler with non-progressive clinical signs. Can. Vet. J. 51, 1007-1010.

Platt, S.R., Chambers, J.N. and Cross, A. 2004. A modified ventral fixation for surgical management of atlantoaxial subluxation in 19 dogs. Vet. Surg. 33, 349-354.

Pujol, E., Bouvy, B., Omana, M., Fortuny, M., Riera, L. and Pujol, P. 2010. Use of the Kishigami Atlantoaxial Tension Band in eight toy breed dogs with atlantoaxial subluxation. Vet. Surg. 39, 35-42.

Reber, K., Burki, A., Vizcaino Reves, N., Stoffel, M., Gendron, K., Ferguson, S.J. and Forterre, F. 2013. Biomechanical evaluation of the stabilizing function of the atlantoaxial ligaments under shear loading: a canine cadaveric study. Vet. Surg. 42, 918-923.

Riedinger, B., Burki, A., Stahl, C., Howard, J. and Forterre, F. 2015. Biomechanical Evaluation of the Stabilizing Function of Three Atlantoaxial Implants Under Shear Loading: A Canine Cadaveric Study. Vet. Surg. 44, 957-963.

Schulz, K.S., Waldron, D.R. and Fahie, M. 1997. Application of ventral pins and polymethylmethacrylate for the management of atlantoaxial instability: results in nine dogs. Vet. Surg. 26, 317-325.

Stigen, O., Aleksandersen, M., Sorby, R. and Jorgensen, H.J. 2013. Acute non-ambulatory tetraparesis with absence of the dens in two large breed dogs: case reports with a radiographic study of relatives. Acta. Vet. Scand. 55, 31.

Thomas, W.B., Sorjonen, D.C. and Simpson, S.T. 1991. Surgical management of atlantoaxial subluxation in 23 dogs. Vet. Surg. 20, 409-412.