Surgical Stabilization of a Craniocervical Junction Abnormality With Atlanto-Occipital Overlapping in a Dog

Abstract: A 3-year-old male neutered Pomeranian presented with severe, poorly localizable pain that was unresponsive to a combination of oral medications (gabapentin, tramadol, prednisone, and methocarbamol) and a fentanyl patch. A Chiari-like malformation with associated syringomyelia was evident on magnetic resonance imaging (MRI). In addition, overlapping of the dorsal arch of C1 and the dorsal aspect of the foramen magnum was suspected from the MRI scans and verified via computed tomography, suggesting a condition similar to basilar invagination/impression in people. At surgery, a combination of foramen magnum decompression with cranioplasty and stabilization of the atlanto-occipital junction was performed. The atlanto-occipital junction was stabilized using an adaptation of a procedure used in people for basilar invagination/impression. Over the next several months, the dog’s clinical signs improved dramatically, allowing substantial reduction of oral pain medications. This is the first report of surgical stabilization for atlanto-occipital overlapping in a dog.

Craniocervical junction abnormalities in humans include a wide spectrum of disorders involving the caudal occipital region of the skull and the C1 (atlas) and C2 (axis) vertebrae.1–3 Reports of nontraumatic craniovertebral junction abnormalities in dogs are primarily restricted to Chiari-like malformations and atlantoaxial instability.4,5 The imaging findings of the dog in the following case report were previously described in a recent case series.6 There are other reports of dogs with Chiari-like malformations that exhibited concurrent abnormalities of the atlantoaxial region.7–9 It is becoming increasingly apparent that, similar to craniovertebral junction abnormalities in humans, there may be several anatomic variants of such abnormalities in dogs. In particular, a disease process referred to as basilar invagination or basilar impression in people describes a situation in which the atlas and/or axis (typically both) moves toward the foramen magnum, leading to clinical signs of neurologic dysfunction.1,3,10 This disorder can occur as a sole abnormality or in conjunction with Chiari type I malformation and atlantoaxial instability.2,10,11

We have reported the magnetic resonance imaging (MRI) and computed tomography (CT) findings of a group of dogs with a disorder atlanto-occipital overlap syndrome when it involves the occiput and C1.6 There are several reports of surgical management of Chiari-like malformations in dogs12–14; however, there have been no reports describing the surgical management of atlanto-occipital overlapping in the dog. The purpose of this case report is to describe the application of a surgical stabilization procedure for manage-
Case Report: Surgical Stabilization with Atlanto-Occipital Overlapping in a Dog

Midsagittal T2-weighted MRI scan of the caudal brain and cervical/cranial thoracic spine demonstrating the lack of cerebrospinal fluid signal at the craniocervical junction (arrow) and the presence of syringomyelia at the level of the third cervical vertebra (C3) and the cranial thoracic spine (inset at level of T1).

FIGURE 1

Three-dimensional CT reconstruction images demonstrating the abnormal location of the dorsal arch of C1 within the foramen magnum.

FIGURE 2

Diagnosis
On presentation, the dog had no neurologic deficits, but it repetitively cried out in apparent pain and attempted to bite the examiner during physical and neurologic examinations, especially when touched anywhere near the spine. A Chiari-like malformation with associated cervicothoracic syringomyelia was suspected from the MRI scans (FIGURE 1). Although difficult to appreciate on the images, it also appeared that the dorsal arch of C1 was protruding into the foramen magnum.

To better ascertain the bony anatomy of the craniocervical junction, a computed tomography (CT) scan of the region was conducted. The patient was premedicated with midazolam (0.4 mg/kg IM) and dexmedetomidine (2 μg/kg IM), induced with thiopental (1.4 mg/kg IV), intubated, and maintained on a constant-rate infusion (CRI) of fentanyl (0.2 μg/kg/min IV) as well as isoflurane and oxygen during the CT procedure. The CT findings were consistent with atlanto-occipital overlapping (FIGURE 2). In addition, the foramen magnum opening appeared abnormally large, with an area of supraoccipital bone apparently missing. To ascertain whether the overlapping of the occiput and C1 was a static or dynamic problem, CT images were procured with the head in a neutral and in a slightly extended position. We attempted to create a “traction” CT view, but the application of cranially directed traction resulted in slight head extension. The overlapping of the occiput and C1 worsened on the extended view, implying a dynamic lesion.
Surgical Procedure

After consulting with the owners regarding medical and surgical options, the decision was made to perform a foramen magnum decompression with stabilization of the atlanto-occipital junction. The following day, the dog was premedicated with midazolam (0.3 mg/kg IV) and pentobarbital (2 mg/kg IV). Intravenous mannitol (1.0 g/kg, over 20 minutes) was administered before induction. Anesthesia was induced with propofol (2.9 mg/kg IV), and the dog was intubated and maintained on sevoflurane and oxygen. Intravenous cefazolin (22 mg/kg) was administered at the time of premedication and repeated q90min during the procedure.

The patient was positioned in sternal recumbency with the neck ventroflexed. The dorsal aspect of the head and neck was shaved and aseptically prepared from the level of the bregma to the level of the fourth cervical vertebra, with a width approximately equal to that of the atlas. A surgical approach for a foramen magnum decompression was performed as described elsewhere.12 Once both the caudal aspect of the occiput and the dorsal arch of C1 were exposed, a foramen magnum decompression was performed using a high-speed air drill and Lempert rongeurs. The preexisting defect in the occipital bone was further enlarged laterally and dorsally, and approximately three-quarters of the length of the dorsal arch of C1 was removed.

A transversely oriented fibrous band of tissue at the cervicomedullary junction was identified and appeared to be causing a constriction of the underlying tissue. This fibrous band, as well as the underlying adherent dura and arachnoid meningeal layers, was incised along a midline plane with a #11 blade. These meningeal layers (dura/arachnoid) and the adhered fibrous connective tissue band were then sharply excised and removed, exposing the cerebellum, caudal medulla, and initial portion of the cervical spinal cord.

After decompression, the patient’s head was relaxed to a neutral position, and five guide holes were made around the circumference of the occipital bone defect using a 1.1-mm drill bit. Self-tapping 6-mm long (1.5-mm diameter) cortical titanium screws were inserted into these guide holes to an approximate depth of 2 to 3 mm. The wings of C1 were exposed, and a titanium screw and 3.2-mm titanium K wire were inserted into the medial aspect of each wing. A length of 3.2-mm titanium K wire was bent in a horseshoe shape and placed around the periphery of the occipital and C1 implants and secured with a ring of polymethyl methacrylate (PMMA). Eight short 3.2-mm K wires were subsequently placed in the PMMA ring, and a titanium mesh/PMMA plate was fashioned and attached to these wires. Closure was routine. A postoperative CT scan was obtained (FIGURE 3). The dog recovered uneventfully from anesthesia.

Follow-Up

After surgery, the patient was treated with a combination of tramadol (3.6 mg/kg q8h), gabapentin (10 mg/kg q8h), and hydromorphone (0.05 mg/kg q4h) for pain management. Intravenous cefazolin was continued (22 mg/kg IV q8h). After 24 hours, the dog vocalized as if in pain when handled, so a ketamine CRI (0.6 mg/kg/hr IV) was instituted, a fentanyl patch (25 μg) was placed, and methocarbamol (46 mg/kg q8h) was administered. The dog was weaned off of hydromorphone and ketamine over the next several days and was sent home on a combination of oral methocarbamol (36 mg/kg q8h), gabapentin (10 mg/kg q8h), and tramadol (3.6 mg/kg q6–12h as needed), in addition to the fentanyl patch.

Approximately 11 days after discharge, the dog presented to the referring veterinarian for an intensification of screaming episodes and sudden loss of appetite. The patient was hospitalized and maintained on a morphine (0.05 to 0.1 mg/kg/hr)/ketamine (5 to 10 μg/kg/min) IV infusion and intermittent injections of hydromorphone (0.05 mg/kg IV). Oral prednisone (0.8 mg/kg q24h) was also instituted, and a fentanyl patch (25 μg) was placed. The dog responded well to treatment and was discharged after 72 hours with the previously prescribed oral medications, in addition to prednisone.

Approximately 7 weeks after surgery, the dog was rechecked at Cornell and did not display any signs of pain. According to the owners, the dog had shown gradual improvement since the brief hospitalization period with the referring veterinarian. Methocarbamol was discontinued at this time, and prednisone was
Case Report: Surgical Stabilization with Atlanto-Occipital Overlapping in a Dog

FIGURE 3

CT images demonstrating placement of the surgical implants.

Postoperative transaxial views.

Three-dimensional reconstructed views.
decreased to an every-other-day schedule (0.8 mg/kg PO q48h). The owners were instructed to administer tramadol on an as-needed basis. At a 5-month postoperative recheck, the dog was doing very well. There was no indication of pain, even on gentle neck palpation. Tramadol had been discontinued 2 weeks previously. Oral prednisone was discontinued at this time, and the patient was receiving oral gabapentin (10 mg/kg q8h) as a sole medication at the time of article submission.

Discussion

In humans, the category of craniocervical junction disorders (also termed craniovertebral junction disorders and occipitocervical instability) includes a large list of congenital, developmental, and acquired conditions affecting the occiput, C1, C2, or combinations of these regions.1–3 The embryologic development of the craniocervical junction is complex and involves an ordered differentiation of multiple sclerotomes that ultimately form the bones of this junctional area. Multiple “mistakes,” probably genetically mediated, can occur during this differentiation, leading to segmentation failure, fusion failure, or combinations of these problems.1 It has been estimated that basilar invagination/impression may be present in 25% to 50% of adult humans with Chiari type I malformation.15 It has been proposed that these concurrent abnormalities of the craniocervical junction may be co-inherited disorders. It has also been proposed that basilar invagination may be the primary disorder, leading to a deformation of the caudal occipital region (i.e., Chiari type I malformation) during skeletal maturation.1,10,15

The veterinary literature contains multiple reports describing malformations of the caudal occipital region in dogs, as well as numerous descriptions of abnormalities of the atlantoaxial region in dogs.4,5 However, only a few reports describe concurrent disorders of the caudal occipital region and the first two cervical vertebrae in this species.6–9 In a recent study,6 it was found that Cavalier King Charles spaniels (the breed most predisposed to Chiari-like malformation) had distinctive atlantoaxial vertebral morphology that differed from that of other dog breeds; however, no association was found between this different anatomy and the development of syringomyelia. In another study,16 absent or miniscule frontal sinuses in small-breed dogs were significantly associated with the presence of syringomyelia. It is becoming increasingly apparent that multiple concurrent developmental abnormalities of the skull and first two cervical vertebrae may be present in dogs with craniocervical junction disorders, as occurs in people. We have often found it difficult to discern on MRI scans the exact bony structure or structures involved in the constrictive effect at the craniocervical junction in cases of “Chiari-like” malformation. Because this can also be a dilemma in humans with craniocervical junction disorders, it is often recommended to conduct both MRI and CT on such patients.1,17

The main purpose of this case report is to describe the adaptation of a surgical procedure used in the management of people with combined Chiari type I malformation and basilar invagination to a dog with apparent Chiari-like malformation and atlanto-occipital overlapping. Basilar invagination of humans most often involves anterior displacement of C2 toward the foramen magnum, which was not apparent in the dog in this report. Because the dog’s disorder did not exactly match that described for basilar invagination, we chose to use the more descriptive term of atlanto-occipital overlapping. We chose to adapt a dorsal (posterior) procedure that allowed foramen magnum decompression combined with rigid fixation of the occiput to the cervical spine. In many human basilar invagination cases, the overlapping and compression are improved with traction based on dynamic CT images, so surgical fixation is performed in traction in such patients.18,19 Our dog’s occiput and C1 were fixed in a neutral or partially flexed position, as this position appeared to alleviate the overlapping that was apparent on the slightly extended head position on preoperative CT images. As this is the first report of its kind, the only valid conclusion that can be reached is that it is possible to apply this surgical method to a small dog with this combination of craniocervical junction disorders. Whether the dog would have done equally well with decompression alone (i.e., standard foramen magnum decompression) or with foramen magnum decompression with a titanium/PMMA plate fixed to the occiput is unknown. It is our impression that surgical intervention...
was successful, judging from the patient’s sustained decrease in signs of pain as analgesic drugs were tapered postoperatively.

The multitude of terms used to describe abnormalities of the cranio cervical junction area in humans is daunting. In contrast, veterinarians tend to limit diagnoses of abnormalities in this region to either Chiari-like malformations or atlantoaxial subluxations. It may be more prudent to use a more encompassing term that includes the subcategories of specific malformations of the occiput and first two cervical vertebrae, such as cranio cervical junction abnormalities, for this group of disorders. The case described in this report demonstrates the importance of thorough presurgical imaging to adequately describe the nature of a cranio cervical junction abnormality in an individual patient. Hopefully, more experience with atlanto-occipital overlapping in dogs will allow more critical evaluation of surgical treatment options in the future.

References