FALLRAPPORT

Craniocervical malformations in a Chihuahua

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Signalment
Chihuahua, 3y2m, male intact, 2.0 kg

Reason for visit
Progressive tetraparesis, ataxia and cervical hyperesthesia.

History
The dog was referred to Evidensia Specialist Animal Hospital Helsingborg due to progressive tetraparesis, ataxia and cervical hyperesthesia since three weeks. The dog had a similar episode two months before that did not respond to treatment with Meloxicam (Metacam 0,1 mg/kg po SID) but resolved with glucocorticoids (Prednisolone 1,25 mg/kg po SID, tapered and ended over three weeks). The dog had no other recorded diseases and was not on any medical treatment. There was no history of any neurological diseases in any close relatives to the dog. The dog was current on vaccination, had tick-treatment with fipronil (Frontline Vet.) and was fed Doggy Professional Mini. In the last year the dog had been visiting Croatia but no other international travels had been made.

Physical/neurological examination
Physical examination was without remarks. The dog was presented with an ambulatory tetraparesis with general proprioceptive ataxia (mainly in the pelvic limbs). Proprioception was decreased in both pelvic limbs (left more than right) and the left thoracic limb. Spinal palpation revealed mild cervical hyperesthesia. Careful ventroflexion, dorsal extension and lateroflexion of the head resulted in mild discomfort. Mentation, behaviour, posture, cranial nerves and spinal reflexes were without remarks.

Neurological localisation
Mainly left sided C1-C5 myelopathy.

Differential diagnoses
Differentials for the dog's clinical signs were degenerative diseases of the vertebral column and adjacent structures (intervertebral disc disease, articular process hypertrophy), inflammatory disorders of the meninges and spinal cord parenchyma (meningomyelitis, MUA), congenital malformations of the vertebral column and the occipital bone (atlantooccipital overlapping syndrome, atlantoaxial subluxation, chiari-like malformation), syringomyelia and spinal arachnoid diverticulum. Based on the signalment, history and clinical findings it was concluded that the top differentials were atlantoaxial subluxation, meningomyelitis, atlantooccipital overlapping syndrome and chiari-like malformation with secondary syringomyelia, even though the other differentials could not be excluded.

Diagnostics
Analysis of CBC and complete serum biochemistry were performed with mildly decreased creatinine, total solids and cholesterol, and a mild increase in sodium (see Table 1). All infectious and inflammatory parameters such as the white blood count and c-reactive protein were normal (see Table 1).

A magnetic resonance imaging (Esaote Vet-MR Grande, 0,25 T) with FSE T2WI sag and tra, SE T1WI tra, STIR dor, 3D HYCE tra and 3D SST1 tra sequences; and post-contrast SE T1WI tra, 3D SST1 dor sequences were obtained of the head and C1-T2. As a contrast agent 0,15 mg/kg of Gadodiamide (Omniscan) was given intravenously. On magnetic resonance imaging (MRI) a dorsal mainly left sided extradural compression of the spinal cord at the atlantoaxial joint could be seen with a small syringomyelia (<1 mm in diameter) caudal to this compression (see Fig. 1 and 2). Occipital hypoplasia and a suspected atlantooccipital overlapping was also seen (see Fig. 1). No other significant lesions or pathological contrast uptake could be found.

To be able to more clearly define the bony structures of the craniocervical junction a computed tomography (CT, Philips Brilliance Big Bore, 16 slice) was obtained in bone tissue reconstructions. Contrast was not used since there was no pathological contrast uptake on MRI and the CT was performed with the suspicion of skeletal malformations.

CT confirmed atlantooccipital overlapping syndrome and telescoping of the dorsal lamina of atlas in to the vertebral canal of axis (see Fig. 3). Comparing MRI and CT, it was concluded that the compression at the atlantoaxial junction was caused by hypertrophic soft tissue which most probably was composed by

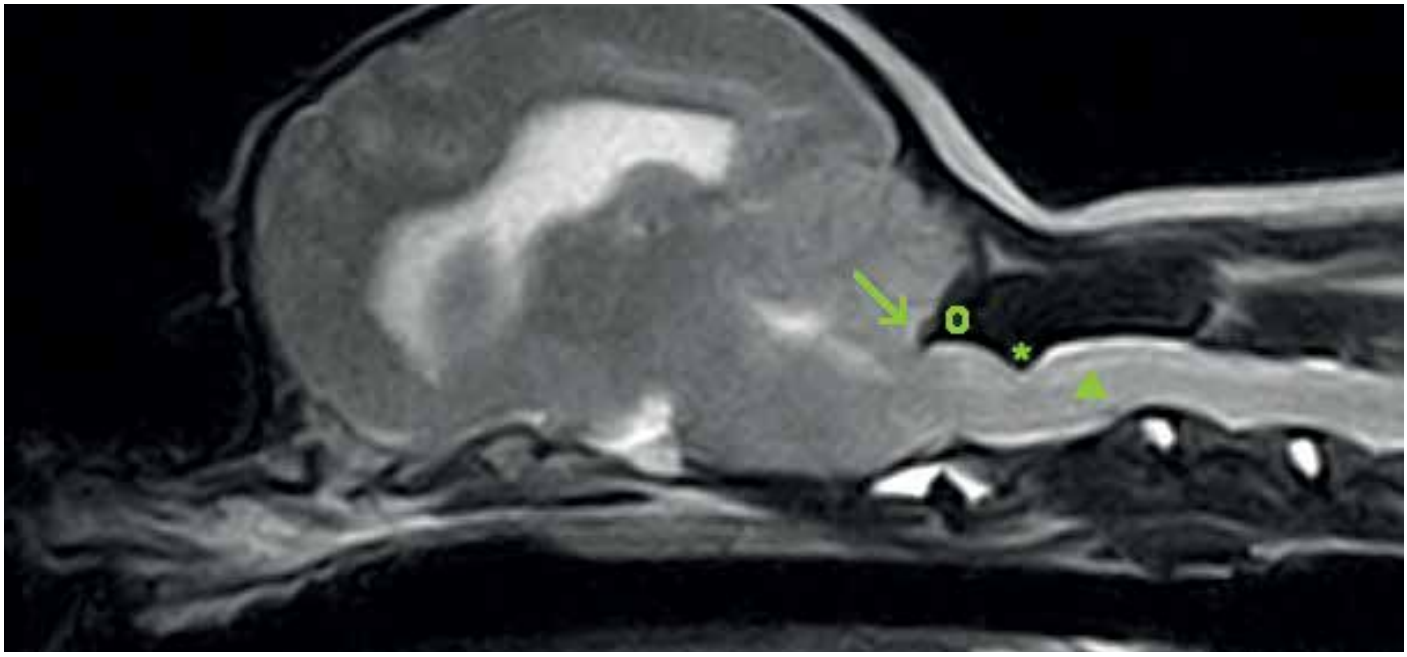

Figure 1. T2WI sagittal orientation of a dog's head and neck. Suspected atlantooccipital overlapping (arrow), dorsal extradural compression of the spinal cord (asterisk), occipital hypoplasia (circle) and a small syringomyelia (arrowhead) can be seen.

hypertrophic interarcuate ligament and/or dura, also called an atlantoaxial dural band. A diagnosis of occipital dysplasia with a dorsally to ventrally widened keyhole-shapen foramen magnum was also made (see Fig. 4).

Diagnosis and prognosis

The patient was diagnosed with several craniocervical malformations including atlantooccipital overlapping syndrome, occipital dysplasia, occipital hypoplasia, an atlantoaxial dural band and telescoping of the dorsal lamina of atlas in to the vertebral canal of axis. A secondary syringomyelia due to the reported craniocervical malformations was also diagnosed. Prognosis was concluded to be guarded.

Treatment

The dog was treated with gabapentin (Gabapentin 10 mg/kg po BID) and a three week tapering course of glucocorticoids (Prednisolone starting on 1 mg/kg po SID). The dog was kept in strict rest at home. Using a harness when walking the dog outside and elevating the food/water bowl were recommended.

Follow-up

By two weeks the clinical signs of the dog had improved. The original clinical signs of the dog were still present but to a lesser degree. By six weeks the dog was clinically and neurologically normal. Treatment with Prednisolone was discontinued after three weeks as planned. The use of Gabapentin (10 mg/kg po BID) and using a harness when walking the dog continued.

Discussion

Atlantoaxial dural bands have been described at the atlantoaxial junction in dogs where it may cause a dorsal extradural compression of the spinal cord (1, 2). It has been observed in conjunction with other abnormalities such as atlantooccipital overlapping and atlantoaxial subluxation (2). A dural band arise

Parameter	Result	Reference range
Hemoglobin (g/dL)	17.1	13.10 – 20.50
Leukocytes (10^9/L)	5.90	5.05 – 16.76
Neutrophils (10^9/L)	4.3	3.00 – 11.50
Band neutrophils (10^9/L)	0	0.00 – 0.30
Eosinophils (10^9/L)	0	0.00 – 1.25
Basophils (10^9/L)	0	0.00 – 0.00
Lymphocytes (10^9/L)	1.1	1.00 – 4.80
Monocytes (10^9/L)	0.5	0.00 – 1.35
Unmature (10^9/L)	0,1	0.00 – 0.00
Nucleated rbc (10^9/L)	0	
Red morphology	Normal	
MCV (fL)	65.1	61.60 – 73.50
MCH (pg)	23.3	21.20 – 25.90
MCHC (g/dL)	35.8	32.00 – 37.90
Hematocrit (%)	47.7	37.30 – 61.70
Trombocytes (10^9/L)	342	150.00 – 400.00
MPV (fL)	9.8	8.70 – 13.20
Sodium (mmol/L)	148	141.00 – 144.00
Potassium (mmol/L)	4.4	4.40 – 5.20
Calcium (mmol/L)	2.5	2.00 – 2.80
ALAT (ukat/L)	0.7	0.30 – 1.30
ALP (ukat/L)	0.2	0.10 – 1.70
Total solids (g/L)	60	66.00 – 75.00
Urea (mmol/L)	2.9	3.80 – 9.00
Creatinine (umol/L)	36	65.00 – 105.00
Cholesterol (mmol/L)	3.6	4.10 – 7.30
Albumin (g/L)	34	29.00 – 39.00
CRP (mg/L)	<10	0.00 – 30.00
Glucose (mmol/L)	3.6	3.80 – 4.90

Table 1. CBC and biochemistry results.

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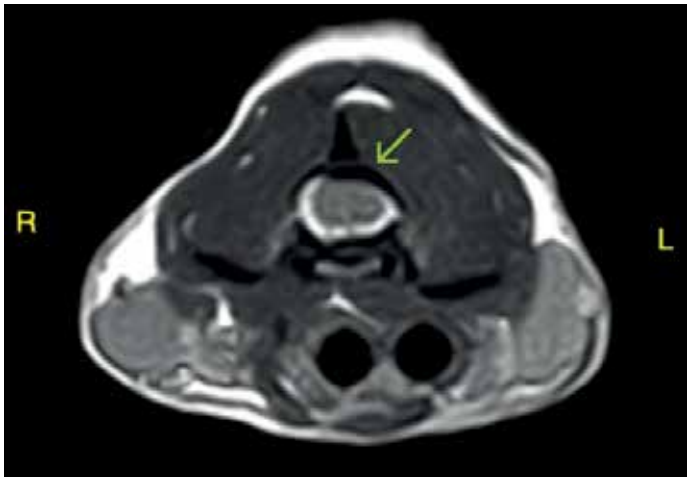

Figure 2. 3D HYCE transverse orientation of a dog's cervical spine at the atlantoaxial joint. A mainly left sided dorsal extradural compression of the spinal cord (arrow) can be seen.

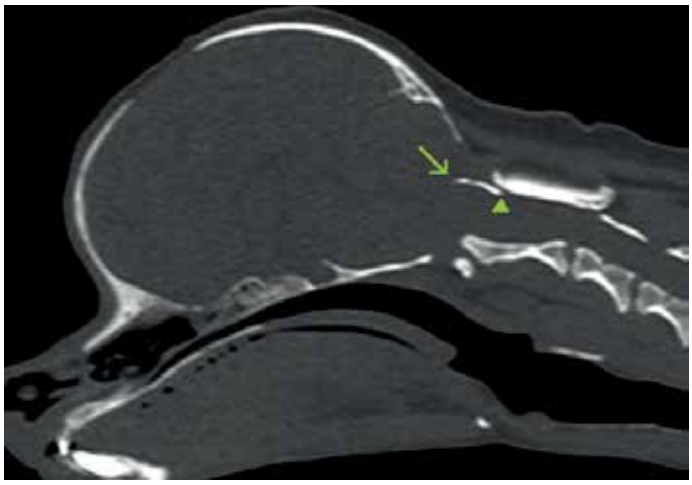

Figure 3. Computed tomography of a dog's skull in bone reconstruction with a sagittal orientation showing atlantooccipital overlapping (arrow) and telescoping of the dorsal lamina of atlas (arrowhead)

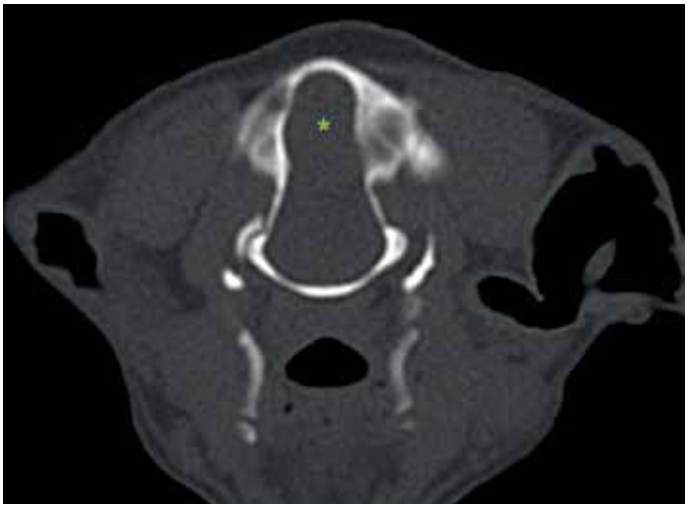

Figure 4. Computed tomography of a dog's occipital bone in bone reconstruction with a transverse orientation showing occipital dysplasia with a keyhole-shapen foramen magnum (asterisk)

from the interarcuate ligament and/or the dura and is composed of inflamed and fibrotic tissue (2). The cause of dural bands is suspected to be a chronic instability of the atlantoaxial joint (1, 2, 3). Atlantoaxial dural bands may cause ataxia, tetraparesis and cervical hyperesthesia (2). Medical treatment includes analgesic drugs and glucocorticoid therapy (2). Surgical treatment is composed by a dorsal laminectomy of the axis with removal of the dural band by sharp dissection (2, 4).

With the MRIs poor bone resolution, it was challenging to conclude definitely what was causing the compression at the atlantoaxial joint. A T1WI sagittal would possibly have given us more information about the origin of the compression, but this sequence was not included in the MRI study. Based on the T2WI sequence the compression could have been caused by a hypertrophic interarcuate ligament, a chronic atlantoaxial subluxation and/or possibly telescoping of atlas dorsal lamina in to axis vertebral canal. Therefore, a CT was performed, which has been recommended by several authors as a complement to MRI when examining the craniocervical junction (1, 2). It can be discussed if there was actual telescoping at the atlantoaxial joint or if there was some other kind of atlantoaxial instability. It is probable that a chronic atlantoaxial instability exists and has caused the development of the atlantoaxial dural band which is suspected to be the main finding in this case.

The dog was also diagnosed with atlantooccipital overlapping syndrome which is mainly recognized in small- and toy-breed dogs (2, 4). Atlantooccipital overlapping is however present in up to 70 % of chihuahuas and may be seen as an incidental finding (5). The malformation causes an overlap between the atlas and occipital bone where the atlas is penetrating the foramen magnum and causes various degrees of compression of the cerebellum and/or the medulla (1, 2, 3). Clinical signs can include neck pain, cervical myelopathy, medullary and/or cerebellar signs depending on the degree of compression (1, 2). The diagnosis is made using CT and/or MRI with a recommendation of using both imaging modalities (1, 2). Medical treatment includes analgesic drugs and glucocorticoid therapy (2, 6). In case of a secondary syringomyelia (1), drugs that decrease CSF-production can be used (2). Surgical treatment consists of foramen magnum decompression with cranioplasty and stabilization of the atlanto-occipital junction (6). Another surgical approach when having both an atlantooccipital overlapping and atlantoaxial instability is standard ventral fixation of the atlantoaxial junction which prevents atlantooccipital overlapping with a recovery rate of 91,7 % (4, 7).

The dog's atlantooccipital overlapping may have attributed to the cervical hyperesthesia but it is unlikely to be severe enough to cause compression of the brainstem. Therefore, the tetraparesis and proprioceptive ataxia is unlikely to be explained by this malformation. The degree of atlantooccipital overlapping, atlantoaxial instability and/or telescoping might however have been underestimated since they may be affected by the degree of flexion/extension of the patient's head/neck during imaging. Dynamic studies may have to be done to fully appreciate the degree of overlapping, instability and/or telescoping (1). Thus, it can not be completely excluded that a dynamic study would not have yielded a different conclusion.

The presence of occipital dysplasia and occipital hypoplasia is also believed to be incidental findings. Occipital dysplasia is common in toy breeds and correlate poorly with clinical signs (1). The malformation may however allow a cranially positioned atlas to displace through the keyhole-shapen foramen magnum causing compression of the cerebellum (1). Occipital hypoplasia

is found in up to 100 % of chihuahuas, with up to 50 % being asymptomatic (5). Reported symptoms related to occipital hypoplasia and secondary syringomyelia in chihuahuas include spinal hyperesthesia and decreased postural reactions (5). Deficits in postural reactions is however significantly associated with a syringomyelia of a minimum diameter of 2 mm at maximal width in a transerverse plane (5). The dog's occipital hypoplasia may be involved in the formation of the syringomyelia, which may explain the cervical hyperesthesia, but the syringomyelia is to small to be a probable explanation for the postural deficits. Future worsening of the syringomyelia and associated symptoms can not be excluded since a combination of occipital dysplasia and occipital hypoplasia may cause less obstruction to CSF flow at the foramen magnum, which may postpone development of a secondary syringomyelia (8).

There were no findings on neither blood analysis or MRI that would indicate that the dog suffered from an inflammatory CNS disease. Even though an inflammatory CNS disease can not be excluded completely (9), the diagnostic results made it unlikely. Ventroflexion of the cervical spine during a cisternal tap would potentially have put the dog under risk for neurological deterioration due to increased spinal cord compression by the atlantoaxial dural band. Thus, a spinal tap was not performed.

The surgical approach for this case would have been a ventral stabilization of the atlantoaxial joint with a dorsal laminectomy of axis to remove the dural band, as reported by Takahashi et al (2018). It is a technically challenging approach which potentially have a high risk for life-threatening complications. The dog was therefore initially put on medical treatment with analgesia and a short course of Prednisolone, which has been described in the literature (2, 6). The dog responded well to this treatment, however a risk of future relapse and neurological deterioration can not be excluded. •

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