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CASE REPORT
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Multiple Malformations of the Craniocervical Junction in German Spitz Adult Dog

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ABSTRACT

***Background*:** Craniocervical junction abnormalities (CJA) are described as complex congenital malformations affecting the caudal occipital region and 1st cervical vertebrae. These abnormalities often include Chiari malformations, atlanto-occipital overlapping, cervical canal stenosis, and atlantoaxial instability. There are few descriptions of these conditions in veterinary literature, despite their relevance. This report's aim is to describe a case of complex craniocervical malformation in a dog, presenting the clinical findings, diagnostic methods, surgical treatment, and post-operative evolution, thus improving the understanding and management of such cases in veterinary practice.

***Case*:** A 4-year-old German Spitz dog, weighing 4.6 kg, was presented to a Veterinary Hospital in São Paulo State with a history of cervical hyperesthesia and non-ambulatory tetraparesis. Neurological examination revealed an absence of conscious proprioception in both thoracic and pelvic limbs, along with segmental hyperreflexia, pointing the neurolocalization of the lesion to the C1-C5 spinal segment. The dog's clinical evolution indicated further involvement of the brainstem, thalamic, and cerebellar regions. Cervical radiographs showed changes suggestive of craniocervical and vertebral malformations at C1-C2. Magnetic resonance imaging (MRI) further elucidated these findings, revealing a complex malformation causing significant compression of the medulla oblongata, atlanto-occipital overlapping, dysplasia/hypoplasia of the occipital bone, and mild cerebellar herniation, leading to partial obstruction of cerebrospinal fluid (CSF) flow. CSF cytology did not show significant abnormalities. Due to the unfavorable evolution of the patient, it was necessary to intervene surgically. The dog underwent foramen magnum decompression (FMD) with suboccipital craniectomy associated to cranioplasty using titanium mesh. Additionally, C1 laminoplasty and C1-C2 laminectomy were performed to maintain the stability of the atlanto-occipital joint and provide access to the dura mater. The dog presented a gradual and complete recovery of its motor function in the post-operative period, indicating a successful surgical treatment.

***Discussion*:** This case highlights the importance of advanced diagnostic imaging, such as MRI, in accurately identifying and assessing craniocervical junction abnormalities in dogs beyond what radiographs are able to identify. The successful surgical outcome obtained in this case shows the effectiveness of combining different surgical approaches for the management of such malformations. The foramen magnum decompression with cranioplasty, C1 laminoplasty, and C1-C2 laminectomy are great examples of fruitful combined techniques. Conservative therapies, such as pain management and the use of carbonic anhydrase inhibitors, may be satisfactory treatment options but could be insufficient for severe cases. Surgical intervention, as demonstrated, can significantly improve prognosis and quality of life by relieving compressive forces on the medulla oblongata and restoring normal CSF flow. The use of cranioplasty is particularly beneficial in reducing complications associated with scar tissue formation. This report adds valuable data to the limited veterinary literature on CJA, emphasizing the need for precise diagnosis and the advantages of the association of surgical approaches. This knowledge will ultimately improve treatment outcomes and the quality of life for animals with such abnormalities.

Keywords: laminectomy, malformation, magnetic resonance, small animals, surgery.

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INTRODUCTION

Craniocervical junction abnormalities (CJA) are described as complex congenital malformations that affect the caudal occipital region and the 1st cervical vertebrae. These are often represented by Chiari malformations (CM), atlanto-occipital overlapping (AOO) and cervical canal stenosis between C1-C2, as well as atlantoaxial instability (AAI) [4].

Young dogs of toy breeds are considered to be the most susceptible to be affected by the condition, with neuropathic pain as the main clinical sign, specifically caused by CM [7,9]. Additionally, cervical hyperesthesia is commonly observed, as well as multifocal neurological signs such as cranial nerves deficits, epileptic episodes, behavioral changes, proprioceptive, vestibular, and/or cerebellar ataxia [7,10].

The magnetic resonance imaging (MRI) and the computed tomography (CT) are the imaging exams used for the diagnosis of CJA [5]. The clinical treatment is based on pain management, with the use of tricyclic analgesics (e.g. pregabalin or amantadine) associated with corticosteroids and carbonic anhydrase inhibitors [7,9]. Although some patients may be treated clinically, JCA are considered surgically treated affections, with the magnum foramen decompression (MFD) being one of the most indicated procedures, possibly along with a cranioplasty with titanium mesh [2-4].

Since CJA are rarely described in the literature, the current case report aims to describing a complex craniocervical malformation case in a German Spitz adult dog with a history of cervical hyperesthesia and non-ambulatory tetraparesis.

CASE

A 4-year-old male German Spitz intact dog, weighing 4.6 kg, was brought to a Veterinary Hospital in the State of São Paulo, Brazil, with cervical and difficulty standing.

The neurological examination revealed a non-ambulatory tetraparesis and severe cervicalgia. Conscious proprioceptive deficits in thoracic and pelvic limbs were noted, as well as hyperreflexia. No cranial nerves deficits or intention tremors were noticed.

According to the neurological exam, the clinical diagnosis of cervical myelopathy was determined, as well as cerebellar and brainstem involvement, and a thalamic lesion, based on changes in thermoregulation. The radiographic examination of the craniovertebral area suggested to craniocervical and vertebral malformations between C1-C2, with a non-compressive or low-compressive dynamic aspect.

Since the radiograph was inconclusive, an MRI was performed to evaluate possible CJA suggested in the x-ray, as well as other causes of cervical myelopathy, such as intervertebral disc diseases. The MRI examination of the brain and cervical spinal cord was performed in transverse T1, T2 and FLAIR sequences, as well as STIR for the cervical area. A complex malformation of the craniocervical region was identified, causing a significant compression of the medulla oblongata with atlanto-occipital overlapping, dysplasia/hypoplasia of the occipital bone and mild cerebellar herniation, resulting in a partial obstruction of the cerebrospinal fluid (CSF) flow (Figure 1). Additionally, a cytological exam of the CSF was performed and no significant changes were found.

According to the clinical signs and the performed examinations were prescribed pregabalin[1] [Pregabalina® - 3 mg/kg, orally, BID] and prednisolone[2] [Prediderm® -1 mg/kg, orally, SID], for a total of 25 days, with a gradual reduction of the prednisolone dosage; manitol[3] [Manitol® - 1 g/kg, intravenous, TID] and furosemide[4] [Furosemida® -1 mg/kg, orally, SID] for 10 days.

The CJA was the clinical diagnosis since the patient didn't show any improvement, and a surgical procedure was decided. During the surgery, it was possible to notice de cerebellar herniation and the dorsoventral compression of the spinal cord at C1-C2. The dorsal aspect of the dura mater between C1-2 was fibrotic, with material consistent with bone fibrosis/metaplasia, causing moderate dorsal compression of the spinal cord on the cranial portion of C2 and dural band sign (Figure 2).

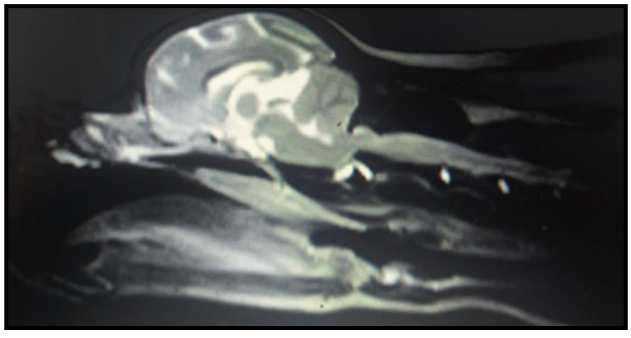

Figure 1. Sagittal T2-weighted MR image, showing a malformation of the craniocervical region and compression of the medulla oblongata with atlanto-occipital overlapping, dysplasia/hypoplasia of the occipital bone and mild cerebellar herniation.

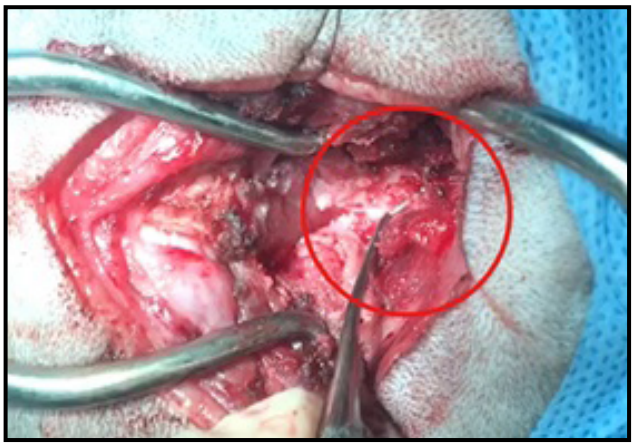

Figure 2. Dural band (red circle) through a medullary exploration.

Since there was a partial obstruction of the CSF flow, ventriculomegaly was observed in the lateral, 3rd and 4th ventricles, and being classified as mild to moderate, and so determining the diagnosis of congenital CJA. The surgical treatment consisted of the correction of the CJA with the MFD being performed through a suboccipital craniectomy combined with a cranioplasty with titanium mesh. The C1 laminoplasty and the right dorsolateral laminectomy between C1-C2 were performed to maintain the atlanto-occipital joint stability, and access to the dural band, which was prominent in the right lateral area. Subsequently, a durotomy to remove the compressive dural band was performed (Figure 3A, B & C).

After the surgical procedure, pregabalin[1] [Pregabalina® - 3 mg/kg, orally, BID] was prescribed for 30 days. The patient improved clinically in the following order: appropriate thermoregulatory response, reduction of cervicalgia, evolution from non-ambulatory tetraparesis to ambulatory tetraparesis in 5 days, and presented gait with little to no ataxia, recovery of proprioception and normalization of segmentary reflexes after 30 days to surgery. After 45 days of the surgical procedure, the patient showed normal spinal reflexes and absence of proprioceptive deficit.

DISCUSSION

The aim of the current case report was to describe the clinical signs, diagnosis, surgical treatment and post-operative evolution of a complex craniocervical malformation in an adult German Spitz dog, since the literature proved to be scarce regarding similar reports. CM is a condition in which there is a flawed development of the cranium and the cervical vertebrae, compromising the accommodation of the encephalon and the cervical spinal cord [9]. Due to the atlanto-occipital instability (AOI), there may be a significant impairment of cerebrospinal fluid flow, leading to new changes, such as syringomyelia and ventriculomegaly, or even the formation of dural bands, which are proliferations of the ligament. This effect may be related to a lymphoplasmacytic inflammatory reaction and bone metaplasia, leading to a dorsal compression of the cervical spinal cord [12].

Alongside the occurrence of CM, often with syringomyelia, the clinical importance of other CJA being associated with toy dog breeds is highlighted, such as AOI and the instability in the craniocervical and atlantoaxial joints. The medullary torsion (or medullary kinking) can also be identified in dogs with CM, resulting in medullary compression [5]. When several CJA are associated, a great clinical and surgical challenge is presented [5]. In the present case report, an initial clinical treatment was conducted which did not result favorably, requiring a surgical intervention. It was necessary to combine several approaches to correct the CJA, elevating the complexity level of the procedure.

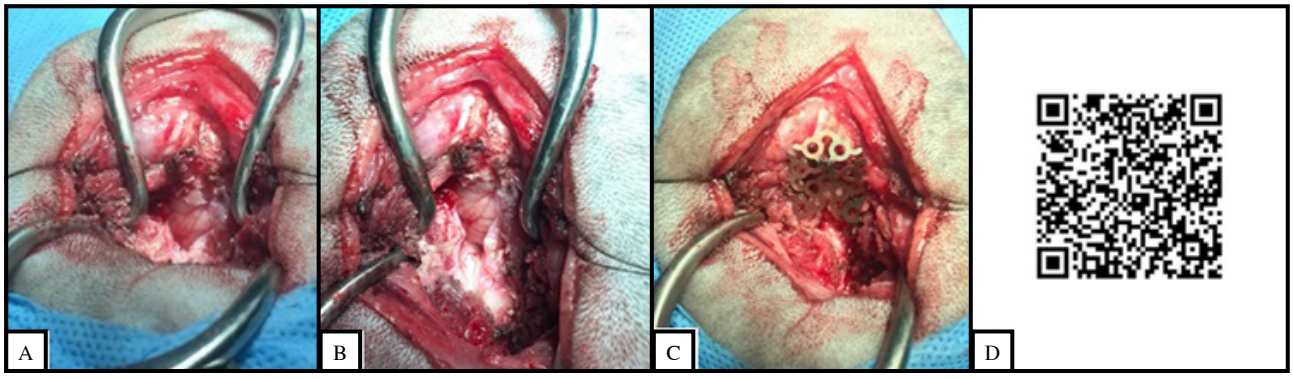

Figure 3. Surgical procedure of the craniocervical junction on an adult German Spitz. A-Suboccipital decompression and dorsocranial laminectomy. B-Final aspect after the decompression. C- Cranioplasty with titanium mesh after suboccipital craniectomy. D- QR code to access videos of the procedure.

Even though the age of occurrence does not represent a reliable parameter for suspecting CJA due to the wide age range of clinical manifestation, the reported patient had 4-year-old and of a toy breed, commonly reported to be predisposed to CJA [4,7]. The age range of dogs suffering from CM ranged from 2 to 4 years on average, with the occurrence mainly in young animals, but may extend by months or years [4,7].

The ventriculomegaly presented by the patient was developed due to an obstruction of the CSF flow by the CJA that affected the craniocervical compliance. The CSF flow obstruction through the foramen magnum, in addition to inducing excessive filling of the ventricles, promotes pressure changes between the blood and the CSF, causing flow alterations that result in the formation of liquid-filled cavities in the spinal cord, known as syringomyelia [7].

A similar case was reported, in which the main clinical sign was tetraparesis and advanced imaging exams were able to elucidate a CM diagnosis associated with ventral displacement of C2, AOI and syringomyelia [6].

Regarding imaging diagnosis, MRI is the gold standard to diagnose soft tissue alterations, which combined with CT, may provide a general overview of the CJA and its complications [1,5]. In the present case, the radiograph was effective in directing the case, since its application enabled the observation of bone alterations suggestive of basioccipital, craniocervical, and vertebral malformations, creating a degree of instability and medullary compression. However, radiographs are not the most appropriate for CJA cases, but may be useful for an initial diagnosis of joint instability and occipital bone dysplasia [11].

In CJA cases, a surgical procedure is indicated due to its higher success rate in the treatment of CM and associated congenital abnormalities. FMD, C1 dorsal laminectomy and ventral fixation of the atlantoaxial joint were performed as surgical treatment, with a satisfactory and positive outcome. The FMD is likely the method of choice in these cases, and is based on a basioccipital craniotomy with a C1 dorsal laminectomy, followed by a durotomy [9]. The cranioplasty may be performed to decrease complications related to scar tissue formation in the area [4,5].

Surgery presents a long-term treatment option for dogs with progressive clinical signs and/or those non-responsive to the conservative clinical treatment. The pain management success rate after surgery may benefit up to 80% of cases [7,8]. The employed surgical techniques followed what was described in the literature, and the cranioplasty was performed using a titanium mesh [7,8].

The craniocervical junction abnormalities can be characterized by several congenital malformations that primarily affect young toy breed dogs. Such malformations may involve various morphophysiological changes, compromising the cerebrospinal fluid flow as well as brain compliance, resulting in significant neurological deficits.

The MRI is the main tool for the diagnosis of craniocervical junction abnormalities, and surgery is the treatment of choice for these abnormalities, especially if the conservative treatment does not satisfactorily stabilize the neurological deficits.

MANUFACTURERS

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REFERENCES

1 **Carrera I., Dennis R., Mellor D.J., Penderis J. & Sullivan M. 2009.** Use of magnetic resonance imaging for morphometric analysis of the caudal cranial fossa in Cavalier King Charles Spaniels. *American Journal of Veterinary Research*. 70(3): 340-345. DOI: 10.2460/ajvr.70.3.340

2 **Dewey C.W., Berg J.M., Barone G., Marino D.J. & Stefanacci J.D. 2005.** Foramen magnum decompression for treatment of caudal occipital malformation syndrome in dogs. *Journal of the American Veterinary Medical Association*. 227(8): 1270-1275. DOI: 10.2460/javma.2005.227.1270

3 **Dewey C.W., Marino D.J., Bailey K.S., Loughin C.A., Barone G., Bolognese P., Milhorat T.H. & Poppe D.J. 2007.** Foramen magnum decompression with cranioplasty for treatment of caudal occipital malformation syndrome in dogs. *Veterinary Surgery*. 36(5): 406-415. DOI: 10.1111/j.1532-950X.2007.00286.x

4 **Dewey C.W., Marino D.J. & Loughin C.A. 2013.** Craniocervical junction abnormalities in dogs. *New Zealand Veterinary Journal*. 61(4): 202-211. DOI: 10.1080/00480169.2013.773851

5 **Hechler A.C. & Moore S.A. 2018.** Understanding and treating Chiari-like malformation and syringomyelia in dogs. *Topics in Companion Animal Medicine*. 33(1): 1-11. DOI: 10.1053/j.tcam.2018.03.002

6 **Itoh H., Itamoto K., Eto S., Haraguchi T., Nishikawa S., Tani K., Itoh Y., Hiyama M., Iseri T., Nakaichi M. & Taura Y. 2017.** Craniocervical junction abnormalities with atlantoaxial subluxation caused by ventral subluxation of C2 in a dog. *Open Veterinary Journal*. 7(1): 65. DOI: 10.4314/ovj.v7i1.10

7 **Loughin C.A. 2016.** Chiari-like malformation. *Veterinary Clinics of North America: Small Animal Practice.* 46(2): 231-242. DOI: 10.1016/j.cvsm.2015.10.002

8 **Rusbridge C. 2020.** New considerations about Chiari-like malformation, syringomyelia and their management. *In Practice*. 42(5): 252-267. DOI: 10.1136/inp.m1869

9 **Rusbridge C., Carruthers H., Dubé M.P., Holmes M. & Jeffery N.D. 2007.** Syringomyelia in Cavalier King Charles Spaniels: the relationship between syrinx dimensions and pain. *Journal of Small Animal Practice*. 48(8): 432-436. DOI: 10.1111/j.1748-5827.2007.00344.x

10 **Rusbridge C., Mcfadyen A.K. & Knowler S.P. 2019.** Behavioral and clinical signs of Chiari-like malformation-associated pain and syringomyelia in Cavalier King Charles Spaniels. *Journal of Veterinary Internal Medicine*. 33(5): 2138-2150. DOI: 10.1111/jvim.15552.

11 **Rusbridge C., Stringer F. & Knowler S.P. 2018.** Clinical application of diagnostic imaging of Chiari-like malformation and syringomyelia. *Frontiers in Veterinary Science*. 5(28): 280. DOI: 10.3389/fvets.2018.00280.

12 **Takahashi F., Kouno S., Yamaguchi S. & Hara Y. 2019.** Evaluation of atlantooccipital overlapping and cerebral ventricle size in dogs with atlantoaxial instability. *Journal of Veterinary Medical Science*. 81(2): 229-236. DOI: 10.1292/jvms.17-0553.