Retrospective study of 16 Cavalier King Charles Spaniel dogs diagnosed with Chiari-Like Malformation and Syringomyelia. Magnetic Resonance Imaging and Surgical Management

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Abstract

Chiari-like malformation (CM) and Syringomyelia (SM) are complex neurological disorders, that affects the brain and respectively the spinal cord. These conditions affect certain toy breed dogs, among which the Cavalier King Charles Spaniel breed is overrepresented, causing numerous clinical signs, of which the neurological pain is commonly observed.

The purpose of this research was to describe the CM/SM magnetic resonance images, to discuss the medical management and to track the outcome of the patients. All of the 16 Cavalier King Charles Spaniel dogs underwent surgical treatment, consisting of craniocervical decompression, combined with drug therapy.

Thirteen of the 16 cases presented good results with clinical improvement of the symptomatology, especially decreasing the neuropathic pain, and increasing the quality of life. The other 3 cases presented no improvement of the symptomatology.

Keywords: Chiari-like malformation, craniocervical decompression, Magnetic resonance imaging.

Introduction

Chiari-like malformation (CM) represents a bony malformation of the cranial caudal fossa, causing herniation of the cerebellum and sometimes the brainstem through the foramen magnum, resulting in cerebrospinal fluid (CSF) flow disturbance. These alterations of the CSF movement may result in the development of fluid filled cavities in the parenchyma of the spinal cord, known as syringomyelia (Rusbridge, 2014). Driver in 2013 in an update study, notes that SM is a progressive, debilitating disease, similar to the same condition in humans.

The Cavalier King Charles Spaniel (CKCS) breed is predisposed to develop CM, due to the

specific juvenile morphology of the skull, and besides that, the volume of their cerebellum compared with other dogs is relatively larger, and this may lead to cerebellar dysfunction. It is estimated that about 92% of the CKCS population is affected by CM (Olsen *et al.*, 2017; Driver *et al.*, 2013).

CM/SM can be encountered also in other toy breeds, such as Griffon Bruxellois, Yorkshire terriers, Maltese terriers, Chihuahuas, Miniature poodles, Bichon Frise, but only in CKCS and Griffon Bruxellois these conditions are suspected to be inherited (Rusbridge 2007).

The aim of this study was to expose the clinical symptomatology of dogs diagnosed with Chiari-

like malformation and syringomyelia, to describe the magnetic resonance imaging results, and to discuss the treatment options and the follow up period.

The most common clinical signs are neuropathic pain, persistent scratching of the neck or shoulder, scoliosis, yelping, proprioceptive deficits and facial paralysis (Rusbridge *et al.*, 2000).

Chiari-like malformation and Syringomyelia are diagnosed based on the brain and spinal magnetic resonance imaging aspects, thanks to the advanced new medical technologies present in veterinary medicine. In the scientific literature several diagnostic imaging techniques of CM/SM are described, including ultrasonography and radiography procedures, and also computed tomography - which is primarily used to perform cranial caudal fossa measurements. Nevertheless, magnetic resonance imaging represents the gold standard in CM/SM diagnosis (Couturier, 2008; Rusbridge, 2009).

British Veterinary Association/Kennel Club Chiari-like malformation and Syringomyelia Health Scheme proposed a grading scale of CM and SM, in order to categorize the conditions. The classification of CM was based on the anatomical position of the cerebellum, thus grade 0 CM represents a dog without CM, in grade 1 CM the cerebellum is indented - mild CM, and grade 2 CM, where the cerebellum is impacted into, or herniated through the foramen magnum. Syringomyelia is divided also in 3 grades, grade 0 SM - normal spinal cord, grade 1- mild SM, the spinal cord presents a central canal dilation or a separate cavity into the spinal parenchyma, with the internal diameter of less than 2 mm, and grade 2 SM, where the central canal dilation has an internal diameter of 2 mm or greater, or a separate syrinx or pre-syrinx with central canal dilation.

Common medications include three main types of medication, drugs used to reduce the cerebrospinal fluid production (furosemide, omeprazole, acetazolamide or cimetidine); analgesics (non-steroidal anti-inflammatory drugs - meloxicam, carprofen, deracoxib, firocoxib; and anti-convulsant drugs that have analgesic properties - gabapentin, pregabalin); and corticosteroids (Rusbridge, 2015; Rusbridge, 2007). Plessas *et al.* in a study performed in 2012 regarding the long-term outcome of Cavalier King Charles Spaniel dogs diagnosed with CM and SM,

found that three-quarters of the dogs presented a deterioration of the neuropathic pain when managed medically.

Motta in a study from 2016 noted that the medical therapy only appears to be ineffective in reducing the size of the syrinx and does not prevent medullary cavity expansion in time. When medical therapy can no longer control CM/SM signs, surgery may be indicated.

The cranio-cervical decompression (CCD) represents the most common surgical procedure used in the management of dogs with CM/SM. This method is also known as suboccipital decompression or foramen magnum decompression, the technique consists in removing the supraoccipital bone and dorsal laminae of the atlas, in order to improve the cerebrospinal fluid flow at the foramen magnum (Rusbridge, 2007). In the literature are described several versions of foramen magnum decompression (FMD), including FMD with a titanium-mesh cranioplasty and durotomy and FMD with durotomy and duraplasty using lyophilized swine submucosa and free adipose tissue autograft implanted over the duroplasty (Ortinu *et al.*, 2015).

Materials and methods

This research was carried out on sixteen Cavalier King Charles dogs, the cases were diagnosed and investigated over a period of 3 years, between 2013 and 2016 at the *PetCode Veterinary Hospital*, Ankara, Turkey.

The selection of the study group was based on the severity and progression of the clinical signs, cases presenting neuropathic pain and with poor response to the drug therapy previously to the CCD surgery. Only dogs that underwent full diagnostic work-up, including a general and neurologic examination, hematology, serum biochemical analysis and magnetic resonance imaging (MRI) scans were considered.

The diagnosis was suspected based on the history, breed, neurological signs and established by magnetic resonance imaging. All cases were evaluated by the same surgeon, and a pain score was assigned to each case, from 0 - no pain, to 4 - severe neuropathic pain, with a serious change in quality of life.

In all cases the magnetic resonance imaging included sagittal and transversal planes of the brain, cervical and thoracic spinal cord, T1- and

T2-weighted scans were performed for each examination.

Due to the various clinical signs, especially the spinal pain, and because of the fact that the brachycephalic dogs are chondrodystrophic breeds, it is important to perform differential diagnosis with Hansen type I disc extrusion, spinal trauma or spinal tumors (da Costa and Moore, 2010).

Cranio-cervical decompression was performed in all cases. Prior to the surgery, pre-medication of the cases was performed with 20 mg/kg intravenous cefazolin sodium (Iespor, I.E. Ulagay), 0.03 mg/kg intravenous medetomidine hydrochloride (Domitor, Orion Pharma, Pfizer Animal Health) and 0.04 mg/kg intravenous atropine sulfate (Atropin 0,2%, Vetaş). Anesthesia was induced with 4 mg/kg intravenous propofol (Propofol 1%, Fresenius Kabi), and maintained with isoflurane 1 – 3 % (Forane, Baxter) in 100% oxygen. Fluid therapy (0,9% sodium chloride injection USP, Baxter) was administered during the entire surgical procedure in constant rate infusion at 5-10 ml/kg/hour, and continued in the postoperative care.

The dogs were positioned in sternal recumbency, on a special operating table, which allows to flex the patient's neck at 60 degrees. The hair was clipped and the cutaneous region of interest was aseptically prepared.

The surgical approach for the cranial cervical decompression was dorsal to the atlas and the supraoccipital bone, cutting the skin and the muscle layers until the bone tissue was exposed. Using a drill and then rongeurs and dental instruments, a supraoccipital craniectomy and a C1 laminectomy was performed. After that, the atlantoccipital membrane was removed, and also all the adhesions present. Dura mater was not sutured, over the laminectomy defect, homologous adipose tissue was placed. Suture of the muscle, subcutaneous and skin layers was routinely carried out.

Postoperatively analgesia was provided with intravenous tramadol hydrochloride, 4 mg/kg (Contramal 100 mg, Abdilbrahim, Grunenthal) at every 8 hours, for 5 days. After the surgical intervention cefazolin was administered in dose of 20 mg/kg, for 7 days, and in order to decrease the neuropathic pain gabapentin was given for 2 weeks, in dose of 10 mg/kg orally, every 8 hours.

The additional long term medical treatment consisted of anticonvulsant drugs, used to relieve the neuropathic pain. Gabapentin was administered in 6 cases, for 2-3 times/ day, in a dose of 10-20 mg/kg, depending on the symptomatology, in order to reduce the neurogenic pain. In 4 cases gabapentin was insufficient, and the clinical signs didn't diminished, thus pregabalin was given in a starting dose of 5 mg/kg up to 10 mg/kg, twice daily. The rest of the cases did not received any long term drugs.

Results and discussions

The study was performed on 16 Cavalier King Charles dogs, with a median age of 35 months, 10 females and 6 males. The youngest patient in the group was 14 months old and the oldest was 97 months old.

Neuropathic pain, spontaneous vocalization and scratching were noticed in all cases, other neurological signs consisted of facial rubbing (50%), air licking (37.5%), paw chewing and licking (37.5%), followed by intermittent head tilt (12.5%), tail chasing(12.5%), circling (12.5%), head tremor (6.25%) and facial paralysis (6.25%).

A neurological pain score was assigned to each patient, according to the neurological pain scale proposed by Rusbridge in a study performed in 2007. The scale is divided in 5 stages, from 0 – no pain, 1 – occasional pain, 2 - mild pain, 3 – moderate pain, to 4 – severe pain. Thus, of all cases 2 dogs matched the pain score 2, 6 cases to pain score 3 and 8 cases to pain score 4.

In all of the dogs included in this study MRI sequences of the brain, cervical and thoracic spinal segments were performed, showing mild indentation of the cerebellum in 3 cases, marked indentation of the cerebellum in 9 cases, and herniation of the cerebellum through foramen magnum in 4 cases. All the cases presented cervical fluid-filled cavities within the spinal cord at the time of examination, the width syrxinx varied between 1.13 mm and 6.73 mm (Table 1).

Wiese *et al.* (2005) highlighted the necessity of the polypharmacology approach, due to the complexity of the neuropathic pain. Corticosteroids have a good outcome for the pain, the disadvantage in performing a long time treatment is that the patients will have immunosuppression, skin, urinary and digestive disorders (Rusbridge, 2007). Therefore for long-term treatment, the

Table 1. Explanatory table of the CKCS dogs included in the study

Case number	Sex	Age(in months)	Magnetic resonance imaging findings	Pain score before the surgery	Pain scale 3 months after the surgery	Long term medication
1.	F	20	Mild indentation of the cerebellum. Cervical width syrinx = 3.81 mm	2	2	none
2.	M	27	mild indentation of the cerebellum. Cervical width syrinx = 2.10 mm	3	2	none
3.	F	22	Marked indentation of the cerebellum Cervical width syrinx = 3.15 mm	4	4	pregabalin
4.	M	48	herniation of the cerebellum through foramen magnum. Cervical width syrinx = 5.56 mm	4	4	pregabalin
5.	M	22	Herniation of the cerebellum through foramen magnum. Cervical width syrinx = 5 mm	4	3	gabapentin
6.	F	34	Herniation of the cerebellum through foramen magnum. Cervical width syrinx = 5.46 mm	4	4	pregabalin
7.	F	22	Marked indentation of the cerebellum. Cervical width syrinx = 1.83 mm	4	3	gabapentin
8.	M	72	Marked indentation of the cerebellum. Cervical width syrinx = 2.18 mm	3	2	gabapentin
9.	M	97	Marked indentation of the cerebellum. cervical width syrinx = 3.40 mm	4	3	pregabalin
10.	F	24	Herniation of the cerebellum through foramen magnum. Cervico-thoracic width syrinx = 6.73 mm	4	3	gabapentin
11.	M	36	Marked indentation of the cerebellum. Cervical width syrinx = 1.13 mm	3	2	none
12.	F	14	Marked indentation of the cerebellum. Cervical width syrinx = 3.28 mm	3	2	none
13.	F	60	Marked indentation of the cerebellum. Cervical width syrinx = 2.14mm	3	2	gabapentin
14.	F	84	Marked indentation of the cerebellum. Cervical width syrinx = 2.80 mm	3	2	none
15.	F	48	Mild indentation of the cerebellum. cervical width syrinx = 1.8 mm	2	2	none
16.	F	84	Marked indentation of the cerebellum. Cervical width syrinx = 4.80 mm	4	3	gabapentin

use of second generation antiepileptic drugs is recommended, such as gabapentin and pregabalin, which may be given concomitantly with non-steroidal anti-inflammatory drugs (Backonja, 2002).

All the dogs made a quick recovery after surgery, the neuropathic pain, spontaneous vocalization and scratching improved in all dogs after the surgery. In the postoperatively period, none of the dogs showed complete resolution of the clinical signs.

The patients were reassessed at 1, 3, 6 and 12 months after the surgery, but a major importance

was the clinical evolution noted by the owner at home. In the follow up period the patients were clinical examined over a period of 1 to 3 years.

At three months post operatively 5 of the 8 dogs with a pain score of 4 had improved to pain score 3; 6 of the cases with a pain score of 3 had improved to 2, and the two dogs with a pain score of 2 maintained unaffected.

Three dogs with a pain score of 4 presented a slightly improvement of the clinical signs immediate after the surgery, but after that showed a gradual worsening of the symptoms at 5, 8 and respectively 12 months after the surgery. Overall

13 dogs presented an improvement of the clinical signs (81%) at three months after the surgery, and all of that 13 cases presented an unchanged evolution in the follow up period. In all this time the owners tracked the evolution of cases at home, adding that the progression was favourable, vocalization and the sudden pain attacks decreased, the scratch behaviour diminished, becoming less frequent and less severe.

The CCD surgical treatment has the role to unblock the CSF pathways at the foramen magnum and thus to reestablish the CSF circulation, improving the pressure and flow, both at cranial and spinal regions. It remains problematic that the results from the current literature show little or no decrease in the syrinx size after surgical management (Ornitau, 2015).

Conclusion

Chiari-like malformation and syringomyelia are complex neurological disorders, difficult to manage, presenting a wide range of clinical signs.

MRI is the gold standard for establish an accurate diagnosis of Chiari like malformation and syringomyelia, providing precise information.

The cervical cranial decompression appears to be effective, improving the clinical symptomatology and the life quality of the patients with CM/SM signs.

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