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SYRINGOSUBARACHNOID SHUNT PLACEMENT IN A CAVALIER KING CHARLES SPANIEL DOG DIAGNOSED WITH CHIARI LIKE MALFORMATION AND SYRINGOMYELIA. CASE REPORT

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Abstract

The aim of this paper was to describe the syringosubarachnoid shunting as surgical management in Chiari like malformation and Syringomyelia, and to track the outcome of the patient. A Cavalier King Charles Spaniel female dog was referred for neurological evaluation due to the intense sudden attacks of pain, occurring more and more often lately. A year earlier, the dog was diagnosed with Chiari-like malformation and syringomyelia, undergoing to a craniocervical decompression with durotomy. Postoperatively the clinical signs improved only for a short period of time, afterwards the clinical symptoms progressively reappeared, worsening in the last two months, thus requiring another surgical intervention. The dog was subjected to surgical placement of a syringosubarachnoid shunt at the cervical spinal cord, the patient being evaluated periodically during the hospitalization, and after that at one, four and twelve months postoperatively. In the follow-up period the patient presented good results with improvement of the clinical signs.
This case report highlights the success of cervical syringosubarachnoid shunt placement in the management of canine Chiari-like malformation with syringomyelia, especially when the other therapeutic options were unsuccessful.

Key words*: Chiari like malformation, dogs, syringomyelia, syringosubarachnoid shunt, magnetic resonance imaging.*

INTRODUCTION

The Cavalier King Charles Spaniel (CKCS) is a small, popular toy dog, originated in the United Kingdom. In Romania the breed presented a grown in popularity in the last ten years, CKCS dogs are sweet-tempered, good with children, playful, and affectionate.
Unfortunately, this breed presents a high predisposition to develop Chiari-like malformation and Syringomyelia.
Chiari like malformation (CM) and syringomyelia (SM) represents an important complex disorder of the nervous system, commonly encountered in the Cavalier King Charles Spaniel dogs, but it can also be diagnosed in other brachycephalic and toy breeds, such as Griffon Bruxellois, Affenpinschers, Yorkshire terriers, Maltese, Chihuahuas, Pomeranians, Boston terriers, Papillons and their crosses (Marino et al., 2012; Rusbridge, 2013).
Canine Chiari like malformation resembles the human Arnold-Chiari type I malformation, a congenital disorder in which the mismatch between the caudal fossa volume, which is too small, and its contents, lead to herniation of the cerebellum and sometimes also the brainstem into or through the foramen magnum (FM), resulting in cerebrospinal fluid flow disturbance (Freeman et al., 2014; Rusbridge, 2007; Rusbridge, 2014).
CM may lead to the development of fluid filled cavities (named also syrinx) within the parenchyma of the spinal cord, condition known as syringomyelia.
Most of the times Chiari-like malformation and syringomyelia evolve together, however, the conditions can evolve independently of the other, affecting dogs aged between 6 months to 10 years old (Rusbridge, 2014; Platt, 2004; Zachary et al., 2011).
The complete mechanism for the development of SM is not fully understood, the most popular theory is that the cavities are formed secondary to the disrupted cerebrospinal fluid (CSF) flow and inconstant CSF pressure (DeLahunta et al.,

2009; Rusbridge, 2014; Cerda-Gonzalez et al., 2009).
The common clinical sign encountered is scratching in the air near the neck - „phantom" scratching, which is the reason why the condition is known as "neck scratcher's disease". Other important sings are the neuropathic pain, expressed by yelping after a sudden posture change, frequent and characteristic manifesttation in CM/SM, along with the head, neck and spinal discomfort (Plessas et al., 2012; Platt, 2004; Rusbridge, 2014; Park et al., 2015)
A variety of non-specific signs accompanies the clinical presentation of CM/SM, including behavioural changes, hyperesthesia, cervical scoliosis, thoracic and pelvic limb ataxia, "bunny-hopping" pelvic limb gait, proprioceptive deficits, vestibular signs and facial nerve paralysis (Rusbridge, 2007; Plessas et al., 2012; Platt et al., 2012).
The gold standard to diagnose CM/SM is magnetic resonance imaging scan (MRI), so that accurate measurements of the caudal cranial cavity can be performed, including the position and shape of the cerebellum, the extension and the symmetry of syringomyelia can also be determined (Platt, 2004; Rusbridge, 2014).
British Veterinary Association (BVA) proposed a CM/SM classification scheme. Based on the position of the cerebellum, CM is structured in 3 grades, from grade 0 - no signs of CM, to grade 2, where the cerebellum is severe herniated through FM.
There is also a grading system for SM, according on the syrinx size there are 3 grades of SM, grade 0 SM – normal spinal cord, to grade 2 SM, where the syrinx diameter is greater than 2 mm (Freeman et al., 2014; Rusbridge, 2014).
The CM/SM grading is important, because it will guide treatment and prognosis.

MATERIALS AND METHODS

A 4-year-old Cavalier King Charles Spaniel female dog was referred for neurological evaluation due to the intense sudden attacks of pain, occurring more and more often lately. The patient was diagnosed a year before with CM and SM, and at that time a craniocervical decompression was performed. After the surgery the patient presented a favorable evolution in the first three months, with the improvement of the clinical signs. Unfortunately, after this period the clinical signs progressively deteriorated over time. In the last two months the dog presented a progressive neuropathic pain, expressed by sudden yelping, neck and spinal discomfort, scoliosis, apathy and depression. Besides these clinical signs, the patient also presented „phantom" scratching and "bunny-hopping" pelvic limb gait.
Due to the medical history and progressive clinical signs, we performed an MRI scan of the brain and spinal cord. Sagittal and transverse T1 and T2 weighted images were acquired. The scan revealed cerebellar herniation, with secondary FM obstruction – corresponding to grade 2 CM (Figure 1).

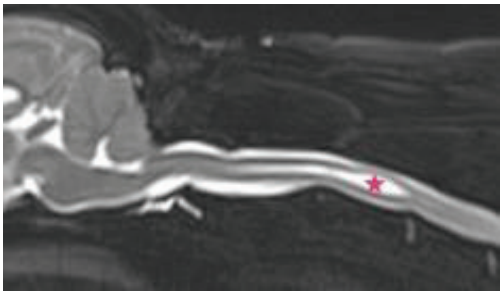

Figure 1 – T2 weighted mid-sagittal image of the caudal cranial fossa and upper cervical spinal cord. The cerebellar vermis is impacted into foramen magnum. Note the syrinx present in the cervical spine (red star).

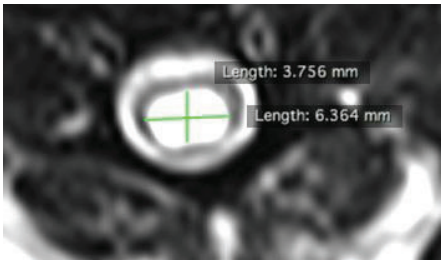

Figure 2 – T2 weighted transverse image of the cervical syrinx. The maximum width of the spinal fluid-filled cavity measures 6.3 mm.

The maximum syrinx width is located cervical, (Figure 1 and Figure 2), marking the position where the shunt will be surgically placed. The width of the syrinx is measured in a transverse plan, in this case the maximum internal diameter is larger than 2 mm, corresponding to grade 2 SM (Figure 2).

The dog was subjected to surgical placement of a syringo-subarachnoid shunt. Prior to the surgery, pre-medication of the dog 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 to 2% (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 10 ml/kg/hour. Postoperatively analgesia was provided with intravenous 4 mg/kg tramadol hydrochloride (Contramal 100 mg, AbdiIbrahim, Grunenthal) at every 12 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 PO orally, every 8 hours.

The dog was positioned in sternal recumbency on a special operating table, which allows to flex the patient's neck at 60 degrees. The cutaneous region of interest is aseptically prepared, the first step of the surgery is to perform dorsal laminectomy at C3-C4 (Figure 3), where the syrinx presents the largest width, previously measured on the magnetic resonance imagining examination.

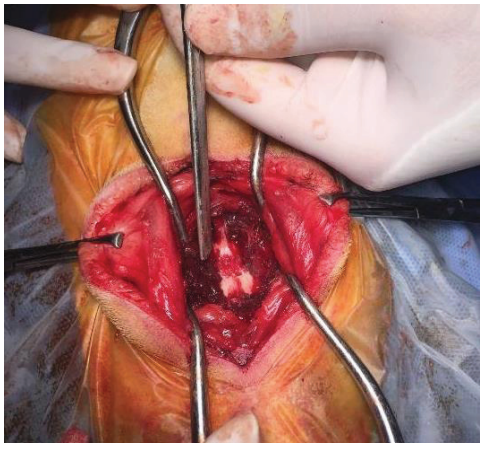

Figure 3 - Intraoperative aspect, performing dorsal cervical laminectomy.

A ventriculoperitoneal catheter was used to create the syringosubarachnoid shunt. Due to the syrinx size, the catheter is tailored to approximately 2,5 cm length (Figure 4), at both terminal ends of the catheter three fenestrations were made, in order to ensure adequately drainage of CSF. One loop is made on the middle of the tube, using nylon thread, having the role of anchoring the tube to the intervertebral ligament.

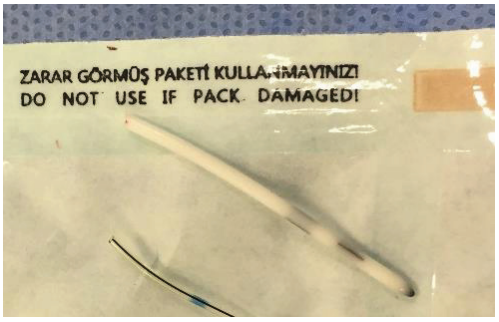

Figure 4 – The catheter used to create the syringosubarachnoid shunt is tailored from a ventriculo-peritonal catheter.

Dura mater and arachnoid are incised with an 11 scalpel blade, presenting an elongated triangular blade, which allows to obtain a thin and precise incision.

The spinal cord is incised in the dorsal midline, penetrating the syrinx. One end of the catheter was introduced into the lumen of the syrinx, in a cranial direction (Figure 5).

The shunt is secured using the nylon loop from the middle of the tube, suturing the shunt to the intervertebral ligament.

The protruding end of the catheter was caudally inserted into the subarachnoid space, thereby achieving the communication between the syrinx lumen and the subarachnoid cavity (Figure 6).

Dura mater was not sutured; homologous adipose tissue was placed over the laminectomy defect. Suture of the muscle, subcutaneous and skin layers was routinely carried out.

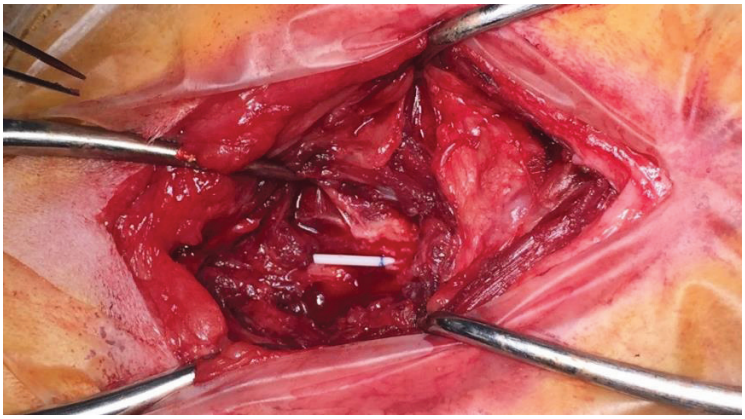

Figure 5 - Intraoperative aspect - one end of the catheter was cranially placed in the spinal cavity. Note the nylon loop present on the middle of the shunt.

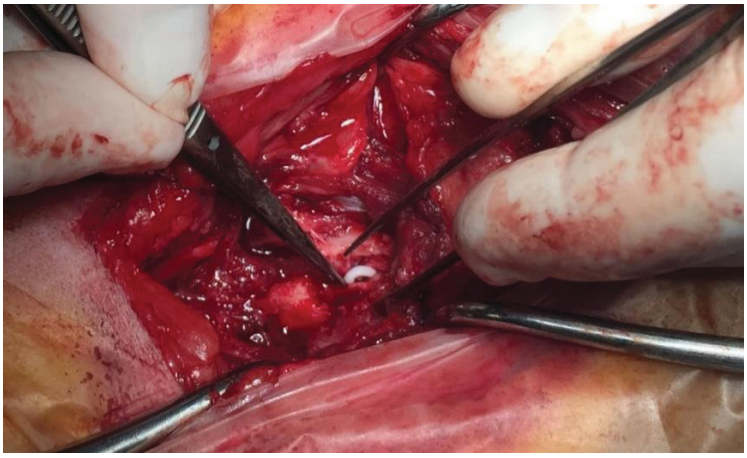

Figure 6 - Intraoperative aspect, orientation of the free-end catheter towards the subarachnoid space, achieving the syringosubarachnoid communication.

RESULTS AND DISCUSSIONS

The MRI scan revealed cerebellar herniation, a wide medullar syrinx located cervical, with the maximum width at the level of the third and fourth cervical vertebrae. In the transverse plan, the maximum internal diameter of the syrinx is 6,36 mm, corresponding to grade 2 SM. Postoperative MRI was not performed.

The syringosubarachnoid shunt was placed at the level of C3-C4, the surgery was successful, no intraoperative or postoperative complications were noted.

We choose to manufacture the tube that we used for shunting from a ventriculo-peritonal catheter, witch fitted optimum to the desired dimensions, and it is flexible and thin. The tube should allow the CSF passage and must be big enough to avoid debris occlusion. In a study performed by Motta in 2012, he chooses to use an equine ocular lavage tube, due to its size, cost and proprieties. The surgical technique that he used resembles, but he did not anchor the shunt to the intervertebral ligament (Motta et al., 2012).

In the early postoperative follow-up period the patient presented a mild neck pain, only for 24 hours. The dog did not presented ataxia, gait abnormalities or proprioceptive placement deficits. After 5 days the dog was discharged, and evaluated after that at 1, 4 and 12 months after the syringosubarachnoid shunt placement.

The neurological examinations in the follow-up period showed progressive improvement of the clinical signs, the neurological pain decreased, and the painful episodes are fewer and have low intensity.

At three years after the syringosubarachnoid shunt placement the patient presents an amelioration of the clinical signs comparative to the preoperative period, notably decreased neuropathic pain, the intensity and the frequency of the pain attacks reduced, scratching and yelping diminished, overall increasing the life quality of the patient.

The main purpose of the treatment is to reduce the pain. There are two treatment options for CM/SM, medical therapy or surgical intervention, which are elected according to clinical symptoms, and can be combined (Rusbridge, 2014).

The most common used surgical technique is craniocervical decompression, which can provide a satisfactory quality of life for a period of time between a few months and two years postoperatively, according to the CM/SM grade and also to the clinical signs. Another surgery technique described is syringosubarachnoid shunting procedure, with a better outcome for patients. Syringosubarachnoid shunt placement can be an option with good results, especially for those patients in which foramen magnum decompression has failed (Motta et al., 2012; Rusbridge, 2014).

Currently the scientific literature offers more information about craniocervical decompression technique, syringosubarachnoid shunting representing a relatively new procedure used in dogs. In patients that had craniocervical decompression, although the syrinx size still persists after this procedure, clinical amelioration is mostly due to the improvement of the CSF flow (Park et al., 2015; Rusbridge, 2014).

In human medicine better results were noticed in patients that underwent syringosubarachnoid shunt placement compared to the ones that underwent only foramen magna decompression or syringoperitoneal shunting. The syringosubarachnoid shunt placement has favorable results regarding the pain control, this technique is preferred to be used in patients with large syringes (Hida et al., 1995).

CONCLUSIONS

Chiari like malformation and syringomyelia represents a complex and progressive neurologic condition, in whose diagnosis and surgical management the magnetic resonance imagining plays an essential role.

Syringosubarachnoid shunt placement represents a safe and efficient procedure, which leads to clinical improvement of the clinical signs and reestablishes the quality of life.

Syringosubarachnoid shunting can be an alternative option with good results, especially in cases in which foramen magnum decompression has failed.

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