CHIARI-LIKE MALFORMATION AND SYRINGOMYELIA IN DOGS

MALFORMAŢIA DE TIP CHIARI ŞI SIRINGOMIELIA LA CÂINE

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ABSTRACT

Syringomyelia is a complex disorder of the spinal cord characterized by the presence of one or more fluid filled cavities within the spinal cord, which by compressing the adjacent nervous tissue leads to various clinical signs - neuropathic pain, sudden yelping, „phantom scratching", ataxia, vestibular dysfunction. The most common predisposing cause of syringomyelia in dogs is Chiari-like malformation, an anomaly characterized by a small posterior cranial fossa compared to the brain tissue, which often leads to herniation of the cerebellum, and sometimes also the brainstem, into or through the foramen magnum (cranial fossa communicates with the spinal canal through the foramen magnum). In this way, the circulation of cerebrospinal fluid is disrupted, leading to secondary lesions and clinical symptomatology.

Chiari-like malformation represents a condition analogue to Arnold-Chiari malformation in humans.

Chiari-like malformation and syringomyelia it is a common condition in dogs, especially in Cavalier King Charles Spaniel dogs and brachycephalic toy breeds.

Cavalier King Charles Spaniel breed is a relatively new introduced in Romania, with a recently increase in popularity. Unfortunately, this breed has a high predisposition to develop Chiari-like malformation and Syringomyelia, a severe neurological condition, more frequent diagnosed lately, thanks to the new generation imaging instruments, such as computer tomography and magnetic resonance imaging.

Chiari-like malformation and syringomyelia are a topic of real interest in canine neuropathology, being a subject of current research due to the fact that in the evolution of the disease are multiple unknowns,and due the fact that currently there is no effective treatment.

Keywords: syringomyelia, Chiari-like malformation, Cavalier King Charles Spaniel, brachycephalic

REZUMAT

Siringomielia reprezintă o afecţiune complexă a măduvei spinării, caracterizată de prezenţa uneia sau a mai multor cavităţi medulare, care prin compresia ţesutului nervos adiacent duce la apariţia simptomatologiei clinice variate – durere neuropatică, plâns/ţipăt din senin, psudo-scărpinat, ataxie, sindrom vestibular. Cea mai comună cauză determinantă a siringomieliei la câine este malformaţia de tip Chiari, anomalie caracterizată prin volumul redus al fosei craniene posterioare în comparaţie cu ţesutul nervos, ce are ca urmare hernierea cerebelului, deseori şi a trunchiului cerebral, prin foramen magnum (comunicarea fosei craniene cu canalul spinal). În acest fel circulaţia lichidului cefalorahidian este perturbată, apărând leziunile secundare şi simptomatologia clinică.

Malformaţia de tip Chiari este o afecţiune analogă malformaţiei Arnold-Chiari din medicina umană.

Siringomielia şi malformaţia de tip Chiari apar frecvent la câinii din rasa Cavalier King Charles Spaniel, cât şi la unele rase brahicefalice de tipul toy.

În România, rasa de câini Cavalier King Charles Spaniel este relativ nou introdusă, iar popularitatea acesteia este în continuă creştere în ultimul timp. Din păcate, această rasă are o predispoziţie crescută a malformaţiei de tipul Chiari şi a siringomieliei, afecţiuni grave neurologice, fiind din ce în ce mai frecvent diagnosticate, graţie instrumentelor imagistice de nouă generaţie, aşa cum este tomografia computerizată, cât mai ales prin imagistica obţinută prin rezonanţă magnetică nucleară. Malformaţia de tip Chiari şi siringomielia reprezintă un subiect de actualitate şi de real interes al neuropatologiei canine, reprezentând tema a numeroase cercetări, datorită numeroaselor necunoscute ale evoluţiei afecţiunii, cât şi a faptului că în momentul de faţă nu există un tratament eficace.

Cuvinte cheie: Siringomielia, malformaţia de tip Chiari, Cavalier King Charles Spaniel, brahicefalie

Syringomyelia (SM) is a condition characterized by the development of fluid filled cavities (named also syrinx) within the parenchyma of the spinal cord, secondary to cerebrospinal fluid (CSF) movement obstruction. One of the most common causes for SM is Chiari-like malformation (CM), a condition characterized by herniation of the cerebellum and sometimes also the brainstem into or through the foramen magnum (FM), due to the mismatch in volume between the caudal cranial fossa and the brain tissue (6, 19, 20).

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Most of the times, Chiari-like malformation and syringomyelia evolve together, nonetheless, the affections can evolve independently of the other. CM may be acute or chronic, affecting dogs aged between 6 months to 10 years old (16).

Although Chiari-like malformation has been described predominantly in Cavalier King Charles Spaniel (CKCS) as a condition that can lead to syringomyelia, the affections have been described in many toy breed dogs, such as Griffon Bruxellois, Yorkshire Terriers, Maltese, Chihuahuas, Pomeranians, Boston Terriers, Papillons, and also in their crosses, particularly to CKCS cross-breed dogs (11, 18, 20).

In CKCS and Griffon Bruxellois dogs the conditions are likely to be inherited (19, 23).

Recent researches has shown that CM can also be observed in cats, more common in brachycephalic varieties, with similar clinical manifestations to those in dogs, but this condition is rarely diagnosed in cats. In 2015, Minato published the clinical and imagistic findings of CM in two cats (13, 20).

Neuroanatomy and pathogenesis

Caudal cranial fossa represents an important landmark of the skull, hosting the cerebellum, medulla oblongata and the pons. From this structure six cranial nerves leaves the cavity. The junction between the spinal cord and medulla oblongata is marked by the foramen magnum, where the CSF flows in both directions, but mostly from the cranial cavity to the spinal cord subarachnoid space (4, 24).

The choroid plexus secretes the cerebrospinal fluid into the ventricular spaces, circulating from the ventricular system in the subarachnoid spaces.

After circulating, the cerebrospinal fluid is drained through arachnoid villi and granulations into the blood, and partly drained along the lymphatic pathways. The main functions of CSF are protection of the central nervous system (CNS) from different traumas, supplies nutrients to CNS and removes waste products from cerebral metabolism (1, 4, 24).

There are multiple theories about the pathogenesis of syringomyelia secondary to Chiari-like malformation, however, the complete mechanism is not fully understood, being the subject of intense research in recent years (20). An underdeveloped caudal cranial fossa leads to a small volume of the fossa, while the brain structures are normal in size.

This anomaly causes herniation of the cerebellum with the obstruction of the CSF flow through the FM.

The cerebrospinal fluid flow is disrupted and CSF pressure is inconstant (2, 4).

The CSF flow disturbances and the fluctuation in pressures seems to present an important role in the development of syringomyelia. Is important to note that the tubular cavites formed in the spinal cord parenchyma, which does not involve the central canal, are defined as syringomyelia (syrinx), while hydromyelia refers to a dilatation in the central canal of spinal cord. These two cavities can communicate to each other, but it is difficult to highlight this communication (4, 25). In early stages, the cavities are microscopically localized dorsal and lateral to the central medullary canal, in the central gray matter. Around the cavites the adjacent medullary tissue is edematous (12).

Most often CM causes secondary lesions such as syringomyelia and hydromyelia, which can be referred to as syringohydromyelia. Besides these lesions internal hydrocephalus, spina bifida or meningomyelocele may rarely be observed (4, 12).

There are multiple theories also about the fluid that fills the medullar cavites in SM, one of the most popular is that the fluid is not CSF, resembles with CSF, but has a lower protein concentration. The fluid that fills the cavities is likely to be extracellular fluid, as a result to the abnormal pressure variations in the spinal cord. Another theory is that the fluid is a mix between the LCR and extracellular fluid (4, 8, 9, 19).

Clinical signs

The predominant clinical feature of SM/CM is the neuropathic pain, along with the head, neck and spinal discomfort. Other clinical signs include behavioral changes, hyperesthesia, cervical scoliosis, thoracic and pelvic limb ataxia, paresis, vestibular dysfunction, facial paresis. Pain is observed in 35% of the affected dogs, expressed by allodynia (pain caused by a non-painful stimulus) and dysaestesia (spontaneous discomfort sensation) (16, 17, 19).

Allodynia and dysaesthesia are manifested by „phantom" scratching, face/ear rubbing, yelping after a sudden posture change, frequent and characteristic manifestations in CM/SM.

„Phantom" scratching or „air guitar" scratching is a unilateral type of scratching, performed with one of the posterior limbs, the limb is in the air directed toward the neck, but the dog does not make skin contact during scratching (16, 17, 20).

The clinical signs are directly correlated with the syrinx location, dimension and symmetry; thereby a dog that presents a narrow and symmetrical syrinx can be clinically asymptomatic, while a dog with an

asymmetric syrinx my show discomfort and neuropathic pain (20). Strangely, there are odd cases in which neuroimaging results revealed the presence of CM and SM, but the dog did not show any clinical signs. It is believed that the incidence of Chiari-like malformation and syringomyelia in asymptomatic population of Cavalier King Charles spaniel is relatively high, in a study performed by Couturier in 2008 on normal CKCS dogs, out of the 16 dogs included in the study, seven had syringomyelia (3, 7).

Dogs with CM commonly presents extropia – ventrolateral strabismus, an outward deviation of the eye (Fig. 1). Although there is no evident connection between CM and epilepsy, in one study, 32% of the dogs diagnosed with CM presented also seizures, and in another study performed on CKCS dogs free of seizures, 12,5 % of the cases developed seizures during the follow up period (10, 17, 20).

Fig. 1. Outward deviation of the left eye in a CKCS. Exotropia is a common sign in dogs with Chiari-like malformation.

Diagnostic

The disease can be suspicioned based on the medical history and clinical signs, corroborated with the breed. The gold standard to diagnose these conditions is MRI, the only disadvantage is for the clinically silent cases, which can remain undiagnosed. Using the MRI the extension and the symmetry of SM can be determined (16, 20).

A common lesion observed in CM is the dilatation of the entire ventricular system, indended or herniated cerebellum (Fig. 2); occipital dysplasia may also be seen. The presence of cavities that containing fluid are an indicator for SM. Usually the most severely affected segments are the cranial cervical (Fig. 2) and cranial thoracic (Fig. 3) segments, however, it is recommended to examine the entire spinal cord, the lesion may be located in any medullar segment (4, 6, 20).

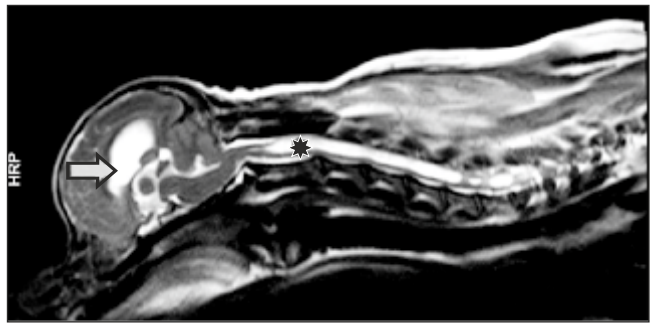

Fig. 2. Midline sagittal T2 weighted MRI image of the brain and cervical spinal cord reveals mild ventriculomegaly (arrow), cerebellar herniation and syringomyelia (star). Normal spinal cord is grey, fluid filled cavites are white.

Among the methods of diagnosis, computed tomography scan is very helpful, the cranial caudal fossa dimensions can easily be measured using this method. Radiography and ultrasonography are also described in literature as useful imagining tools, but their limitations are quite large.

The only method to confirm the CM/SM presence is using the magnetic resonance imaging (8, 21, 22).

The syrinx width is a strong indicator of neuropathic pain, on which the SM grading system is based on.

There are 3 types of SM, starting from grade 0 SM, which is normal, to grade 2 SM, where the syrinx diameter is greater than 2 mm (Table 1).

The diameter of fluid filled cavities must be measured in transverse plane. Another lesion that can be observed in SM is spinal cord edema, an increased fluid content area, considered to be a boundary lesion prior to syrinx formation, named pre-syrinx (6, 20).

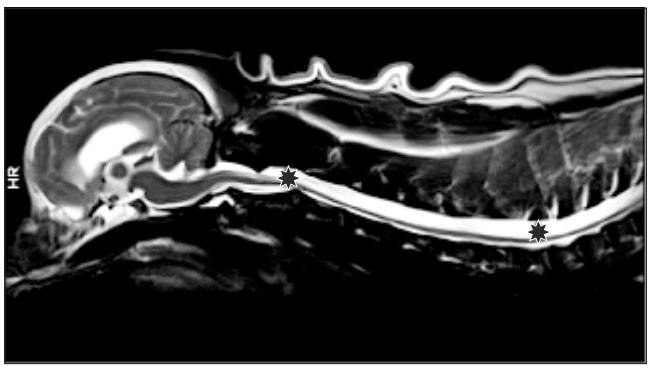

Fig. 3. Midline sagittal T2 weighted MRI image of a 3 year old CKCS dog presenting severe neuropathic pain. Note the severe syringomyelia affecting the cervical and cranial thoracic segments of the spinal cord (stars). Normal spinal cord is grey, fluid filled cavites are white.

There is also a grading system for CM, structured

in 3 grades, according to the position of the cerebellum; from grade 0 - no signs of CM, to grade 2, where the cerebellum is herniated through FM (6, 20, 26) (Table 1).

Table 1

Grading system of Chiari-like malformation and Syringomyelia

Condition / Grade	Chiari-like malformation	Syringomyelia
Grade 0	No Chiari malformation	Normal spinal cord
Grade 1	Indented cerebellum	Central canal dilation or a separate syrinx, with an internal diameter value less than 2 mm or a pre-syrinx without central canal dilation
Grade 2	Impacted or herniated cerebellum into foramen magnum	Hydromyelia with an internal diameter value equal or more than 2 mm, a separate syrinx, or a pre-syrinx with central canal dilation

The differential diagnosis must be performed with other causes of pain and spinal cord dysfunction such as intervertebral disc disease, granulomatous meningoencephalomyelitis, atlantoaxial subluxation, neoplasia and discospondylitis (20).

Treatment and Prognosis

The most important aspect of therapy is to reduce the neuropathic pain. There are 2 types of therapeutic options, medical and surgical, which should be chosen based on clinical symptoms.

In asymptomatic dogs or in dogs with mild non-progressive signs, the treatment may not be necessary, requiring only the support of close supervision. (16, 20)

Medical therapy includes 3 categories of drugs: analgesics (NSAID and anti-epileptic drugs with analgesic properties), corticosteroids and drugs that reduce CSF formation (acetazolamide, furosemide, cimetidine, omeprazole). Usually the initial medical therapy starts by using the analgesics and drugs that reduce CSF production. A good management of neuropathic pain is performed with Gabapentin, 10 mg/kg PO BID, or Pregabalin, 5 mg/kg PO BID. (16, 20)

It is important to note that fully recover is rarely, approximately 70% of patients will show some improvement. (16, 20)

If medical therapy alone is not enough to keep under control the clinical signs, the surgical decompression of the foramen magnum should be made, performing suboccipital craniectomy. The success rates for this option it's relatively high, around 80%. Usually the drug therapy should also be maintained, mainly due to the fact that syrinx size usually persists after the surgery is performed. (16)

Another type of surgical management with favorable results in syringomyelia is placing a syringosubarachnoid shunt. In order to create the shunt, a veterinary ocular lavage catheter (Five French equine ocular lavage catheter) is placed in the syrinx cavity, after performing a dorsal laminectomy, depending on the syrinx location, frequently in the cervical or lumbar spinal cord. This method can lead to a reduction in size of the syrinx, and secondary improvement of the clinical signs. (14)

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