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Review Article

Understanding and Treating Chiari-like Malformation and Syringomyelia in Dogs

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Chiari-like malformation (CM) and syringomyelia (SM) are common and debilitating conditions in toy and small breed dogs. CM, considered ubiquitous in the cavalier King Charles spaniel (CKCS) population, results in abnormal cerebrospinal fluid dynamics which can lead to the development of SM. The clinical signs associated with CM/SM are frequently confused with other otologic and dermatologic conditions, which may delay appropriate treatment. A definitive diagnosis of CM/SM requires advanced imaging; however, due to expense associated with this, many cases are managed presumptively and symptomatically for the condition. The primary goal of treatment is to manage neuropathic pain and neurologic deficits through pharmaceutical or surgical approaches. Current literature suggests that most CM/SM-affected dogs have progression of their clinical signs in spite of medical or surgical management; however, most maintain a good quality of life based on owner assessments. Lack of correlation between structural markers of disease and clinician and owner-derived measures of neuropathic pain highlight the need for more robust, quantitative measures of pain in this common veterinary disease.

Introduction

Chiari-like malformation (CM) and syringomyelia (SM) are common conditions in small and toy breed dogs such as the Cavalier King Charles Spaniel (CKCS) and can result in debilitating clinical signs. CM is a complex malformation that results in caudal fossa crowding and displacement of the cerebellum into the foramen magnum. Resultant craniocervical junction overcrowding causes abnormal cerebrospinal fluid (CSF) dynamics, leading to the formation of SM (fluid cavities within the spinal cord). CM is universally documented in the CKCS population, and SM has been diagnosed in up to 70% of CKCS.[1,2] The clinical signs associated with CM/SM can resemble dermatologic or otologic conditions, presenting a diagnostic quandary for veterinarians evaluating CM/SM-affected dogs. It is important for small animal practitioners to be familiar with common clinical signs of the disease in order to facilitate early intervention and appropriate therapy. The goal of this article is to provide a comprehensive review of the veterinary literature with respect to CM/SM to enhance the companion animal practitioner's understanding of this common disease in order to aid in recognition, diagnosis, and treatment.

Understanding the Pathogenesis

CM and SM are 2 conditions affecting the craniocervical junction that often coexist in certain small and toy breeds such as the Cavalier King Charles Spaniel, Brussels Griffon, Chihuahua, and others. CM is defined as a decrease in the volume of the caudal fossa that results in caudal displacement of the cerebellum, and occasionally brainstem, into or through the foramen magnum.[3] CM is referred to throughout the veterinary literature by various names including caudal occipital malformation syndrome (COMS), occipital hypoplasia, Chiari malformation, and hindbrain herniation. Recently, the Chiari-like Malformation and Syringomyelia Working Group round table designated, by consensus, that the condition should be referred to as Chiari-like malformation (abbreviated CM) in the context of the canine condition.[3] CM has recently been recognized as a breed-specific characteristic, with an incidence of 100% in the CKCS.[2] Approximately 50%-70% of dogs with CM develop SM, though not all show clinical signs associated with the condition.[1,4] CM/SM is typified by a complexity of malformations of the calvarium and occipital bone which result in crowding and in some cases caudal displacement of the cerebellum (CM) as well as fluid accumulation within the spinal cord parenchyma, termed SM. (Fig 1).

Although well-documented in people, the importance of CM in the absence of SM as a cause of clinical signs in dogs is unclear. Some dogs display behaviors suggestive of neuropathic pain when CM is present as a sole abnormality[5,6]; however, CM is essentially universal in the CKCS breed, with many dogs asymptomatic for the condition.[2]

Syringomyelia is directly translated from Latin as "cavity within the spinal cord." The term was introduced by Ollivier d'Anders in 1827 to describe what was ultimately confirmed to be the normal spinal cord central canal.[7,8] The term has since been used to describe a variety of intramedullary cystic structures; however, SM is currently defined within the veterinary literature as the development of fluid-containing cavities within the parenchyma of the spinal cord resulting from abnormal CSF movement.[3] This single term is now used to encompass both of the previously distinct entities of syringomyelia and hydromyelia. A singular accumulation of fluid is termed a "syrinx" and the plural of syrinx is "syringes."

The composition and origin of the fluid within a syrinx is not well defined; however, recent studies in people support the notion that it may not be of CSF origin but rather composed of extracellular fluid. Several studies have compared the protein of CSF to

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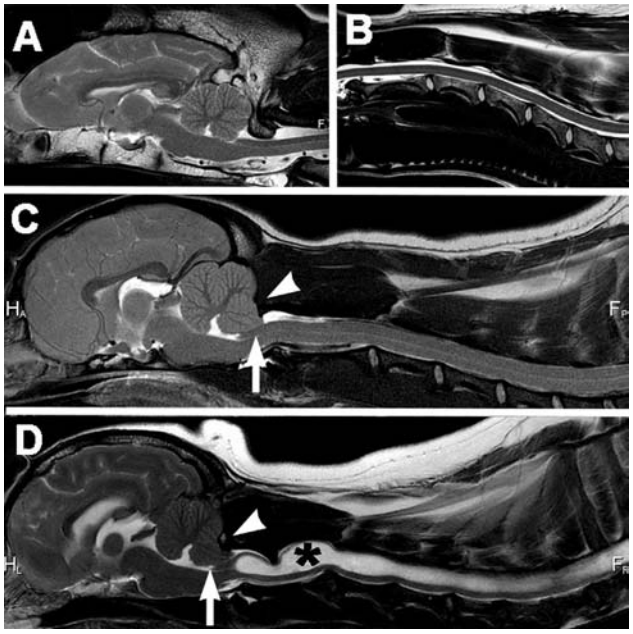

Fig. 1. T2-weighted sagittal images of brain and cervical spinal cord from a normal mesaticephalic dog (A and B), a Cavalier King Charles Spaniel (CKCS) with Chiari-like malformation (CM) but no evidence of syringomyelia (SM; C), and a CKCS with both CM and SM (D). Note the malformation of the caudal aspect of the occiput (C and D; arrow head) and caudal displacement of the cerebellum into the foramen magnum (C and D; arrow) typical of CM. T2 hyperintensities consistent with dilation of the central canal or fluid accumulation within the spinal cord parenchyma are typical findings in dogs with SM (D; asterisk).

that of syrinx fluid with mixed results.[9-13] One study identified a higher protein concentration in the posttraumatic syrinx, 0.35-3.9 g/L, when compared to CSF, 0.1-0.44 g/L, while others found indistinguishable protein concentrations.[9,10]

Although SM is more classically associated with clinical signs, dogs with SM can also be apparently asymptomatic for their condition.[1,14] This suggests that the development of clinical signs with both CM and SM may be multifactorial or that certain disease-modifying elements may play a role.[6,15-17] Given the apparent complexities in the relationship between CM, SM, and the development of clinical signs, a wealth of recent veterinary studies have focused on evaluating the genetic, structural, and biomechanical contributors to this syndrome. Of primary focus related to the pathogenesis of CM/SM has been skull morphology, presence of concurrent anomalies, and alterations in CSF flow dynamics.

Skull Morphology/Morphometry

Given the high prevalence of both CM and SM in the CKCS, numerous studies have evaluated breed-specific skull morphometry in an attempt to better understand its contribution to the development of SM. A summary of recent morphometric studies, their results, and implications for the development of SM are summarized in Table 1. Brachycephalicism has been investigated as an underlying cause of SM due to the lack of published cases of CM/SM in mesaticephalic and dolicocephalic breeds. Brachycephalic dog breeds have an earlier spheno-occipital synchondrosis (growth plate) closure compared to mesaticephalic breeds, with CKCS's having an even early closure.[18] The spheno-occipital synchondrosis is responsible for longitudinal growth of the skull,[19] and early spheno-occipital synchondrosis closure is the putative cause of the short but wide skull shape of CKCS.[18,20] Numerous morphometric studies have explored the correlations between various aspects of the malformation and the development of SM, but to date, a single morphometric characteristic has not been implicated as the primary factor.[6,21,22]

Presence of Concurrent Anomalies

Medullary kinking is a structural anomaly often identified in dogs with CM/SM and consists of elevation of the medulla from the ventral surface of the calvarium at the cervicomedullary junction independent of external boney compression.[34-36,40] This term was adopted from the human literature and is seen in 70% of people with CM.[41] Medullary kinking has a reported prevalence of 66%-100% in CKCS with CM,[26,35,42] and a higher degree of medullary kinking has been associated with increased likelihood of clinical signs associated with CM/SM.[36]

CKCS with CM/SM can have concurrent ventriculomegaly that is more severe than what is typically present in dogs with CM alone. This finding may be a reflection of more significant CSF flow disturbances at the craniocervical junction.[29] Previous reports have suggested that enlarged ventricles secondary to CM may result in seizures.[4] This theory was contradicted in a later study that found no correlation between the presence of seizures and CCF overcrowding or ventriculomegaly.[43] It is the authors' perspective that idiopathic epilepsy, and not a complication of CM/SM, is a more likely cause of seizures in an otherwise healthy, young CKCS with only CM/SM and ventriculomegaly on MRI and a normal CSF analysis.

Several recent studies have highlighted the clinical importance of other concurrent craniocervical junction abnormalities (CJA) beyond CM/SM in toy breed dogs. These anomalous conditions can occur in isolation or in conjunction with CM/SM and include conditions such as atlanto-occipital overlap (AOO),[32,34] dorsal atlantoaxial bands,[35,44,45] and congenital dorsal angulation of the dens.[26,35,46] When present in dogs with CM/SM, these conditions may exacerbate clinical signs and contribute to the worsening of SM.[33] They may also present clinically relevant hurdles, or at least special considerations, in surgical correction of CM/SM; however, an in-depth discussion of other CJAs is considered beyond the scope of this article, and we refer the reader to other excellent recent reviews focusing on other CJAs.[40,47,48]

Cerebrospinal Fluid Flow

Cerebrospinal Fluid (CSF) makes up 10% of the intracranial fluid volume[49,50] and is produced at a relatively constant rate, regardless of intracranial pressure, by the transport of water, potassium, chloride, and bicarbonate from the choroid into the ventricles.[50] The process of CSF production is facilitated by carbonic anhydrase C, sodium and potassium ATPases, and aquaporins in the epithelial lining of the choroid plexus.[50] Aquaporin-4, the most abundant aquaporin found in the brain and spinal cord, is responsible for water transport within the central nervous system and alterations in function of this protein have major implications on CNS homeostasis.[51]

The pathway of CSF flow starts in the lateral ventricles, courses through the interventricular foramen, and into the third ventricle. From there it passes through the mesencephalic aqueduct into the fourth ventricle. The majority of CSF passes into the subarachnoid space through the lateral apertures adjacent to the foramen magnum; however, a small fraction also passes through the central canal.[52] CSF flow rate, pressure, and drainage are influenced by body position, physical activities, gravity, blood pressure, and accelerating forces.[52-54] Alterations in CSF flow have been implicated as a major contributing factor to the development of SM, and there are several theories as to how this may occur.

The "water hammer theory" was first described in 1950 by Drs. James Gardner and Robert Goodall, 2 physicians who reasoned