Syringomyelia: An emerging spinal disease in dogs

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Spinal cord diseases are common in dogs. With the advent of magnetic resonance imaging, diseases that occur exclusively or predominantly within the spinal cord are more readily diagnosed compared to previous years. Of these 'newer' intraspinal diseases, the most common are associated with abnormal intraspinal cord fluid accumulations consistent with syringomyelia and hydromyelia. Possible pathogenic mechanisms of syringo-/hydromyelia formation include changes in CSF pressure, relationships within the spinal cord (as occurs with hydrocephalus or foramen magnum abnormalities), loss or abnormal development (myelodysplasia) of spinal parenchyma, stenosis of the central canal and obstruction to CSF flow from inflammation or tumours. The recent recognition that syringomelia and hydromyelia are common conditions in dogs has resulted in an increased interest in the pathophysiology associated with these spinal diseases as well as a search for treatment options for affected animals.

Introduction

Numerous diseases can result in signs of spinal cord dysfunction (LeCouteur *et al*., 1995). Included in this group of diseases are those, such as intervertebral disc disease, that compress or distort the cord from an extradural location and those diseases that arise from within the spinal cord parenchyma itself. With the advent of newer spinal imaging modalities, the ability to 'see' inside the spinal cord has greatly improved. Consequently, diseases that occur within the spinal cord itself are being diagnosed with increasing frequency.

Syringomyelia and hydromyelia are prominent examples of intraspinal cord diseases that were previously thought to be rare, but are now commonly diagnosed in animals with spinal cord dysfunction (McGrath, 1965; Bone and Wilson, 1982; deLahunta *et al.*, 1983; Child *et al*., 1986; Cauzinille and Kornegay, 1992; Johnson *et al.*, 1992; Bagley *et al.*, 1996; Chauvet *et al.*, 1996; Rusbridge *et al.*, 2000; Tage *et al.*, 2000). A citation search with the key word 'syringomyelia' in both humans and animals yields over 2,000 citations since the 1960s, suggesting that this is not a new disease process. The prevalence, however, was thought to be low until very recently. With the use of better spinal imaging modalities for diagnosis of spinal disease, these cystic spinal cord diseases are now identified as being surprisingly common.

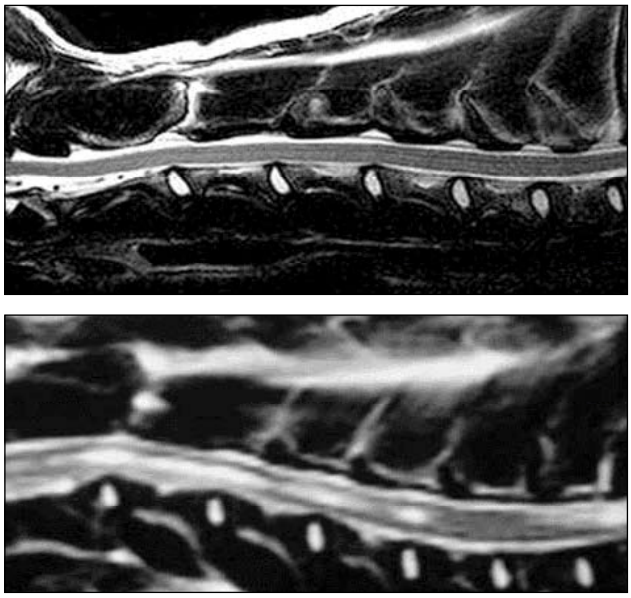

Figure 1: Sagittal T2-weighted MR image of the cervical spine from a clinically healthy dog (A, top) and a dog with syringomyelia (B, above).There is significant increased signal (hyperintensity; whiter) consistent with excessive fluid accumulation in the spinal cord (arrows).

Syringomyelia and hydromyelia are essentially cystic abnormalities of the spinal cord. Syringomyelia refers to abnormal cavities filled with liquid in the substance of the spinal cord (McGrath, 1965); a 'syrinx' is the term used to describe one of these pathological cavities. Hydromyelia refers to a condition characterised by the accumulation of fluid within an enlarged central canal of the spinal cord. In both of these instances, the fluid that accumulates is similar, if not identical, to cerebrospinal fluid (CSF). The term syringomyelia is often used to describe all of these types of intraspinal abnormal fluid accumulations (Milhorat *et al.*, 1995).

Clinically and diagnostically it is often difficult to differentiate between syringomyelia and hydromyelia. Ultimately, the diagnosis is made when the affected spinal cord is evaluated histologically. A hydromyelic cavity is lined by ependymal cells characteristic of the central canal and a syringomyelic cavity is found within the spinal cord external to the central canal and is lined by glial cells. In some instances, the differentiation between the two is difficult as an enlarging hydromyelia may destroy or disrupt the ependyma layer of the central canal resulting in fluid escaping into the surrounding spinal cord. Alternatively, an enlarging syringomyelic cavity may expand into the central canal resulting in hydromyelia (Milhorat *et al.*, 1995).

Pathologic mechanisms that result in syringomyelia and hydromyelia are complex and variable, with no single pathogenic mechanism adequately explaining all nuances of the diseases (Milhorat *et al.*, 1995). Possible pathogenic

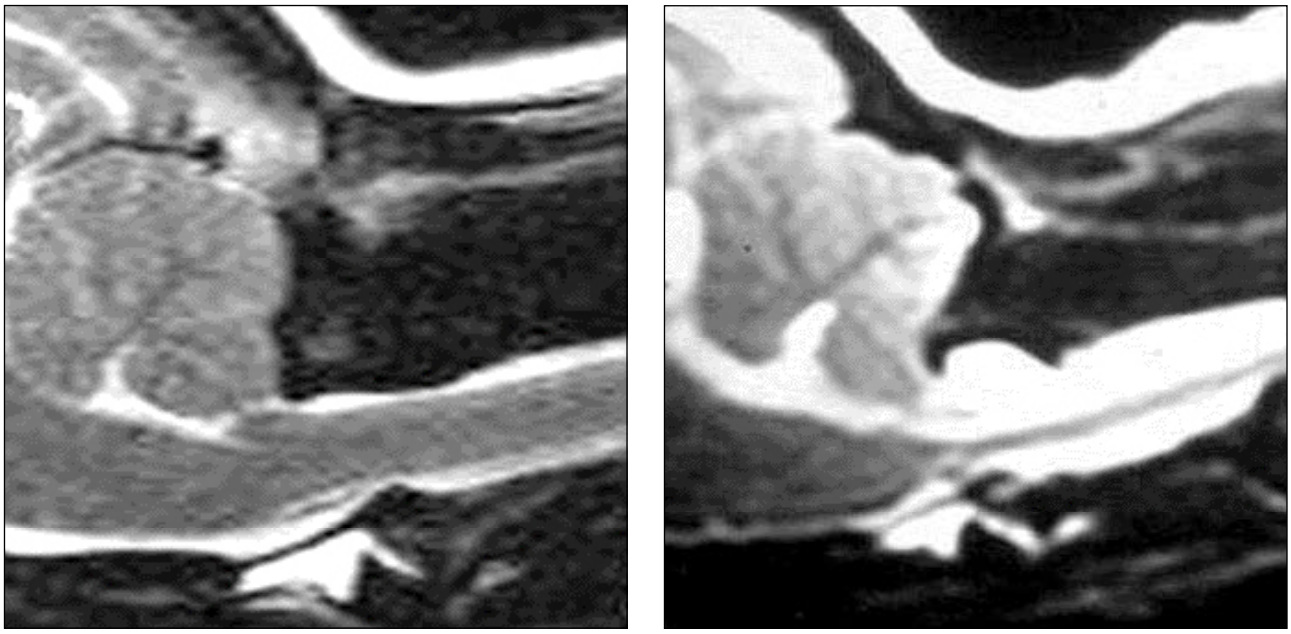

Figure 2: Sagittal T2-weighted MR image from the foramen magnum region of a clinically healthy dog (A, left) and a dog with syringomyelia (B, right).

mechanisms of syrinx formation include changes in CSF pressure relationships within the spinal cord (as occurs with hydrocephalus or foramen magnum abnormalities), loss or abnormal development of spinal parenchyma (myelodysplasia), stenosis of the central canal and obstruction to CSF flow via inflammation or tumor.

Syrinxes that do not communicate with the central canal (extracanalicular syrinxes) at any level are often acquired due to spinal injury or damage from disease (haemorrhage and inflammation) (Milhorat *et al.*, 1995). These cavities tend to occur in the central grey matter or dorsal and lateral white matter, possibly associated with changes in vascular distribution ('watershed zones') (Oi *et al.*, 1991). Larger defects, often extending the majority of the length of the spinal cord, are associated with congenital defects or diseases acquired early in life.

Hydromyelia is often associated with hydrocephalus in humans and has been reported rarely in dogs (Hall *et al.*, 1980; Oi *et al.*, 1991; Milhorat *et al.*, 1995; Itoh *et al.*, 1996). In many instances of syringomyelia, however, abnormalities exist at the foramen magnum that ultimately result in syringomyelia.

Studies of humans with posterior fossa abnormalities (referred to as Chiari malformations) have shown that disequilibration and movement of CSF from the intracranial to the spinal subarachnoid space may be the underlying factor in perpetuating syringomyelia. If the foramen magnum is obstructed due to caudal displacement of the cerebellum, CSF cannot move in either direction. Cerebrospinal fluid which cannot leave the intracranial space during systole causes increased intracranial pressure. The pulsatile increase in pressure is transmitted down the spinal cord and appears to be an important factor in perpetuating the syrinx cavity. It has been shown previously, in normal dogs, that there are CSF pulsations concurrent with the heartbeat, and that there is an increase in CSF pressures from the caudal to cranial cervical area (Bagley *et al.*, 2000). Also, using a similar pressure recording system, it has been shown that intracranial pressures are significantly higher than cervical spinal cord pressures. These data combined would indirectly suggest that there may normally be a pressure differential (step-down) at the foramen magnum area.

Congenital and acquired defects of the foramen magnum area are a group of diseases that have been described in animals, some with similarities to the Chiari-type malformations in humans. This group of diseases is primarily associated with extension of the caudal cerebellum dorsal to the medulla oblongata and caudal to the foramen magnum around the cervical spinal cord and displacement of the medulla oblongata and fourth ventricle into the cervical spinal canal. These abnormalities were originally grouped into four separate entities by Chiari in the late 1800s and are sometimes refereed to as 'Arnold-Chiari malformations.' These malformations are frequently associated with hydrocephalus and syringo/hydromyelias are commonly encountered concurrently. Obstruction of CSF flow at the foramen magnum seems to be the primary mechanism of both of these pathologic changes. Similar abnormalities may occur in dogs and other animals.

Clinical features

The clinical signs of syringo/hydromyelia reflect spinal cord dysfunction and reflect the predominant neuroanatomical area of lesion involvement. The clinical signs associated with syringomyelia are rarely pathognomic. Common signs of spinal cord dysfunction with syringomyelia are similar to those seen with other diseases of the spinal cord including

ataxia, paresis and pain. Interestingly, a number of dogs and humans with syringomyelia also have scoliosis (Kokmen *et al.*, 1985; Child *et al.*, 1986; Goldberg and Dowlings, 1991). Bladder and bowel dysfunction may also be found. Some Cavalier King Charles Spaniels affected with syringomyelia in association with a malformation of the caudal aspects of the foramen magnum and brain stem (Chiari malformation) have an unusual facial scratching or rubbing behavior (Rusbridge *et al.*, 2000). Any dog, however, with signs of spinal cord dysfunction at any level within the spinal cord may have syringomyelia and a high index of suspicion for this disease is often necessary for ultimate diagnosis. Syringomyelia tends to be found more often in the smaller breed dogs such as Pomeranians, Chihuahuas, Maltese terriers and Poodles. Cavalier King Charles Spaniels are also commonly affected with this disease, and even Cavalier King Charles Spaniels without clinical signs may have this condition to some degree. Importantly, syringomyelia may be present as a component of, or independent of, other spinal cord diseases such as atlantoaxial subluxation and intervertebral disk disease.Therefore, it is important to have a high index of suspicion for this disease, even when other spinal diseases that are more obviously apparent are present.

Diagnostic testing

The diagnosis of syringomelia can be difficult, as these abnormalities are often not apparent with routine myelography. In rare instances, with lumbar injections of contrast medium during myelography, it's possible to fill the central canal with contrast medium allowing visualisation of hydromyelia. However myelography is frequently normal in affected animals.

Of the other spinal imaging studies available, magnetic resonance (MR) imaging is often the most helpful in establishing a diagnosis (de Groot *et al.*, 1984; Kjos *et al.*, 1985; Kokmen *et al.*, 1985; Elster *et al.*, 1986; Chauvet *et al.*, 1996; Haughton *et al.*, 1999) (**Figures 1** and **2,** pp 35-36). Many syringomyelic lesions contain primarily water and varying degrees of protein and fat. Magnetic resonance images of cystic lesions have long values of T1 and T2, resulting in hypointense and hyperintense signals, respectively. Different image characteristics on MR, however, may be noted with differing consistencies of the fluid present.

With MR imaging, the syrinx cavities may be located in any region of the spinal cord or spinal segment. However, they are more commonly found in the cervical spinal cord segments, involving the dorsal aspect of the spinal cord. The cavities may be discontinuous, and be located multifocally within the spinal cord. The edges of these cavities may be discrete or irregular. When the cavities enlarge, it is often difficult to determine whether there is communication with the central canal or overlying subarachnoid space. In some situations with early syringomyelia, the spinal cord will appear diffusely hyperintense on T2-weighted studies (**Figure 3**). This may represent a diffuse oedematous phase during the early phases of this disease process.

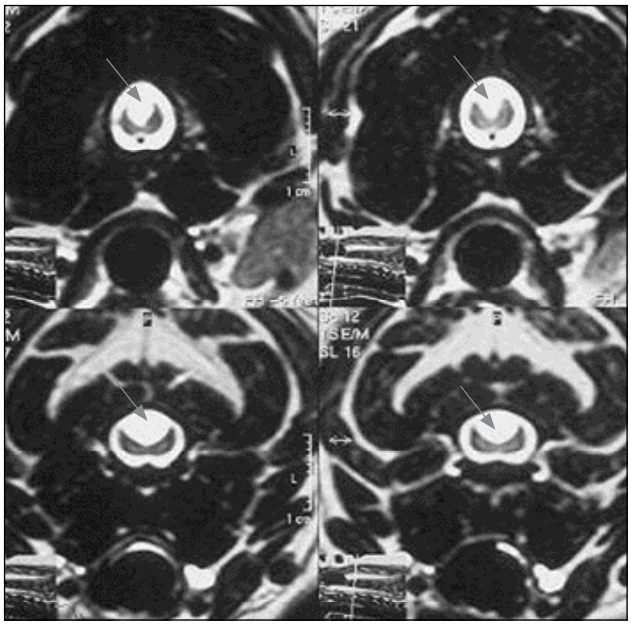

Figure 3: Transverse T2-weighted MR image of a dog with syringomyelia.

As syringomyelia is often associated with abnormalities of the posterior fossa and foramen magnum as well as hydrocephalus, it is important to image the intracranial area in dogs with syringomyelia. In Cavalier King Charles Spaniels, these spinal cord cavities often appear irregular. When these abnormalities are identified with MR, it is also important to assess the caudal brain stem and foramen magnum region for associated anatomical abnormalities. In Cavalier King Charles Spaniels, for example, the brain stem and cerebellum may be abnormally displaced or compressed. In fact, the cerebellum may actually be shifted through the foramen magnum into the dorsal cranial cervical spinal cord region. In this breed there is often an associated abnormal development of the occipital bone, wherein this bone is more 'indented' just rostral to the dorsal foramen magnum opening. This bony 'indentation' often crowds the cerebellum, resulting in the cerebellar displacement. The overall size of this area of the cranial cavity is also often small.

Treatment

In humans, the treatment of cranial cervical syringomyelia, with or without caudal fossae abnormalities, remains controversial. In instances where the lesions are subclinical or clinical signs are mild and non-progressive, no definitive treatment may be needed (Johnson *et al.*, 1992; Tage *et al.*, 2000). Clouding the issue further, some syrinxes have spontaneously regressed, resulting in some authors questioning the role of surgery as treatment for this problem. If clinical signs are progressive, however, more definitive treatment for syringomyelia should be considered. Surgical treatments available include incision (decompression) of the syrinx via myelotomy, syringosubarachnoid shunting, ventriculoperitoneal shunting and posterior fossa decompression via a suboccipital craniectomy and associated cervical vertebral laminectomy. Myelotomy and

cyst decompression may be helpful to relieve pressure within the syrinx cavity. In some syrinxes, however, intralesional pressures are not excessive and myelotomy, therefore, will not be helpful. Shunting of the syrinx cavity may be helpful in selected cases, but post-surgery shunt occlusion is a common problem. Ventriculoperitoneal shunting from the lateral ventricle similarly has resulted in mixed improvements.

For isolated fourth ventricular dilation, fourth ventricular shunting has been performed in humans and dogs. The shunt is placed from the fourth ventricle into the peritoneal cavity. Technically, placement of the proximal catheter tip into the fourth ventricle may result in iatrogenic trauma to the brain stem. Morbidity as high as 42% have been associated with placement of the rostral catheter tip in humans (Lee *et al.*, 1995). Clinical signs include cranial nerve palsies (of cranial nerves XI, VII, and VIII), head tilt, paresis and ataxia.

In instances where foramen magnum abnormalities exist, which result in alterations in CSF pressures, removal of any stenosis or resistance to CSF flow may be helpful. This type of posterior fossa decompression in dogs is performed via a suboccipital craniectomy and may require a partial laminectomy of C1. If occipital dysplasia is present, a fibrous covering is present in the caudal occipital bone area. This tissue is incised with a scalpel blade or microscissors. If the occipital bone is intact, a high-speed nitrogen-powered drill is useful in removing the occipital bone. Similar to performance of a laminectomy, the bone is removed with the drill to a thin layer of inner cortical bone ('egg-shell thin'). The remaining bone is removed with rongeurs. The more fibrous dura is then incised with a scalpel or scissors. Cerebrospinal fluid flows freely from the area of the dura incision. Prior to closure, a dura substitute is placed over the craniectomy and laminectomy defect. A fascial graft collected from the temporalis muscle can be used for this purpose.

Treatment responses are influenced by the appropriate diagnosis, an understanding the pathophysiologic events that result in or perpetuate syringo- and hydromyelia, and the type of pathology present. With the increasing use of MR imaging for evaluation of spinal disease, syringomyelia and hydromyelia are being diagnosed with increasing frequency and are, in fact, surprisingly common abnormalities in dogs. Recognition the existence of these diseases should be helpful when considering the differential diagnosis of dogs with spinal cord dysfunction.

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