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 syringomyelia 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.

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
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 disequilibrium 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 referred 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
During the early phases of this disease process, will appear diffusely hyperintense on T2-weighted studies in some situations with early syringomyelia, the spinal cord with the central canal or overlying subarachnoid space. In difficult to determine whether there is communication discrete or irregular. When the cavities enlarge, it is often within the spinal cord. The edges of these cavities may be discontinuous, and be located multifocally any region of the spinal cord or spinal segment. However, they are more commonly found in the cervical spinal cord. 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.

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.

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 ‘indentated’ 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, intrasional 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 is placed over the bone, and the craniectomy defect is closed. A dural graft is then placed over the bone to protect the dura substitute. A dural substitute is then placed over the bone to protect the dura substitute.

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.

References


