Inflammation of the Central Nervous System- Encephalitis, Myelitis, and Meningitis

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Inflammation of the central nervous system is a very common condition seen in veterinary neurology. The area in which the inflammation predominates indicates which of the clinical syndromes we use to name the disease. Inflammation involving the brain is called encephalitis, of the spinal cord is called myelitis and of the meninges is called meningitis. It is common to have more than one area of the central nervous system affected and therefore combinations of the names are often used such as meningoencephalomyelitis or meningoencephalitis. Encephalitis is the most common area of the central nervous system to be affected and will be used as the general term in this article.

Etiology

There are two main classes of encephalitis: infectious and idiopathic. Infectious etiologies include viruses, fungi, bacteria, protozoa, tick borne diseases and even algae. True infectious encephalitis is much less common in the dog than in other species including humans; however diagnosis is critical for proper targeted therapy. Idiopathic encephalitis means that an infectious etiology has not been found and encompasses a wide range of syndromes appreciated in the canine patient. Many idiopathic encephalitidies are thought to be immune-mediated and respond well to immune-suppression. Other idiopathic encephalitis, such as necrotizing encephalitis found in young toy breed dogs, do not appear to respond as well to immuno-suppression and therefore may represent another form of the disease. Idiopathic encephalitis is much more common in the dog than infectious encephalitis; however, definitive diagnosis is critical as treatment is markedly different between the two categories of disease.

Idiopathic Encephalitis

As previously noted, idiopathic encephalitis encompasses a wide range of syndromes including, Granulamotous Meningoencephalitis (GME), Necrotizing meningoencephalitis (NME) and Necrotizing Leukoencephalitis (NLE). There can be overlap of these disease syndromes within the same dog. We are also now recognizing overlap of breeds traditionally thought to only be affected by one disease such as Pug dog Encephalitis or Yorkie Leukoencephalitis. MRI characteristics can be very helpful in differentiating between syndromes in some cases, but histopathology is still the gold standard. It is important to realize that because diagnosis was traditionally based on histopathology; older literature is extremely biased to a poor outcome as diagnosis was based on necropsy. Though encephalitis is still a very serious disease, many dogs can respond favorably to medications and treatment should be attempted in most cases.

Clinical Signs

Clinical signs vary depending on the area of central nervous system affected. Common signs appreciated with encephalitis affecting the forebrain are seizures, blindness, behavior changes, mentation changes and/or circling. When the brainstem is involved, ataxia, head tilt, head turn,

cranial nerve deficits and mentation changes can be seen. The most common clinical signs associated with myelitis are paresis and ataxia and can often look identical to other causes of myelopathy such as intervertebral disc disease. Strictly speaking, meningitis is only inflammation of the meninges and therefore the only clinical signs that are typically present are pain (can be extreme) and sometimes fever. Only one clinical sign such as seizures or a combination of all the above signs can be found depending on how focal or diffuse the inflammation of the central nervous system is. Many cases of encephalitis can be rapidly progressive where others can have seizures as the only clinical sign for months prior to diagnosis.

Diagnosis

Infectious encephalitis can affect any dog at any age and variables include environment, vaccine history and immune competency. Granulamatous Meningoencephalitis is generally a disease of middle age dogs and tends to affect toy breeds, however large dogs can also get GME. Necrotizing Meningoencephaltitis is most commonly seen in young Pugs, Maltese, Chihuahua, Boston Terriers, and Shih Tzus. Necrotizing Leukoencephalitis is most commonly seen in young Yorkshire Terriers and Chihuahuas. Since seizures are a common clinical sign of encephalitis, it should be high on the differential list for any young toy breed dog that starts having seizures and further diagnostics should be considered in these breeds.

Cerebrospinal fluid (CSF) analysis is the most useful diagnostic test for inflammatory disease of the CNS; however, inflammation can also occur with some tumors and even intervertebral disc disease. Because of this, in many cases, we combine MRI and a CSF tap to give us the most information. Occasionally a CSF tap can be falsely normal, either because the animal has already been given steroids or because the inflammation is deep in the parenchyma and the CSF is not representative. If the MRI is also suggestive of encephalitis but the CSF analysis is negative, we can often treat presumptively on the MRI information. Because steroids are such a strong anti-inflammatory drug, even a single dose can make a CSF analysis normal in a dog that has encephalitis. It is generally not recommended to give steroids prior to referral if encephalitis is on the differential list, as this can preclude diagnosis.

Once encephalitis is diagnosed, additional infectious disease testing is often performed to rule out an infection. Antibody or antigen titers and PCR performed on CSF and/or blood are the most common tests used to look for potential infections. CSF culture is occasionally performed if there is a high index of suspicion for a bacterial infection.

Prognosis

Prognosis is incredibly variable and highly dependent on the type of encephalitis diagnosed and the speed of diagnosis. Encephalitis is very serious and carries a fair to guarded prognosis in all cases. Infectious encephalitis is often difficult to treat because of the blood brain barrier and lack of penetration of many drugs. Viral encephalitis has limited treatment options and usually entails supportive care. Bacterial infections, most frequently from extension of otitis interna or animal bites, can often be treated very effectively with antibiotics and surgery (bulla osteotomy,

abscess removal, etc.). Fungal infections are notoriously difficult to clear completely and some animals require lifelong therapy with anti-fungal drugs.

Prognosis with idiopathic encephalitis is also highly variable and difficult to predict. Some dogs begin treatment with steroids alone and respond immediately and fully while other require additional immune-suppressant medications. Other dogs improve but may never return to normal function or their signs recur every time medications are tapered or withdrawn and therefore require lifelong medication. Still other dogs, such as those with necrotizing encephalitis, do not tend to respond as well to immune-suppressive medications and often continue to have intractable seizures or other worsening clinical signs that result in death or euthanasia.

Again, though encephalitis is a very serious disease, it can have a good outcome in many cases if a diagnosis is obtained and aggressive treatment is instituted early.