Inflammatory CNS disease

Inflammatory CNS disease is a broad term used to describe a number of conditions causing inflammation of the central nervous system (from here on referred to as CNS).

Depending on which part of the CNS is inflamed, inflammatory CNS disease can be more precisely divided into meningitis (inflammation of the meninges), encephalitis (inflammation of the brain) and myelitis (inflammation of the spinal cord). Each condition can occur on its own, but it is usually combined with at least one of the other two (e.g. meningo-encephalitis, meningo-myelitis).

What are the causes of inflammatory CNS disease?
Causes of inflammatory CNS disease can be either infectious or non-infectious. Infectious causes are probably the least common and can be due to viral (Distemper in dogs, feline-infectious peritonitis, FIV), bacterial, protozoal (Toxoplasma, Neospora) or fungal agents. Non-infectious causes are more common and include breed-specific disorders of Yorkshire terriers, Maltese and Pug dogs as well as the presumed immune-mediated disorder (granulomatous meningo-encephalitis also known as GME).

The later is probably the most common cause of inflammatory CNS disease in dogs. Why the immune system suddenly becomes ‘over-excited’ and decides to inflame the CNS remains a matter of speculation. Other rare non-infectious causes include preneoplastic disorders (inflammation that will turn into cancer with time) and paraneoplastic disorders (cancer outside of the nervous system that has an effect on the nervous system—inflammation, in this case).

What are the signs of inflammatory CNS disease?
The signs of inflammatory CNS disease vary according to which part of the CNS is inflamed (brain, spinal cord and/or meninges).

When meningitis occurs on its own, pain, reluctance to move the neck, a hunched-up back and walking stiffly are the most common signs. Fever is seen in less than half of the cases and its absence cannot be used to discard the possibility of meningitis. Encephalitis and myelitis are associated with neurological signs reflecting which part of the nervous system is inflamed. For example, any or all of the following may be seen with encephalitis—seizures, walking in circles, behavior changes, facial or throat weakness, head tilt and/or falling. Myelitis may be associated with pain, difficulty walking, falling or knuckling on one or all limbs, or being unable to walk at all.

How do you diagnose inflammatory CNS disease?
Unfortunately, the diagnosis of inflammatory CNS disease cannot be based solely on the clinical and/or neurological signs as other neurological conditions (tumour, brain bleed) can potentially cause similar signs. Often all blood work is normal even in the most severe cases of meningitis or encephalitis.
Further tests such as an MRI can re-enforce the suspicion of inflammatory disease and rule out other causes of neurological disease. Cerebrospinal fluid analysis (CSF) collection is probably one of the most useful tests. Not only can CSF help to confirm the presence and type of inflammation but important tests can be carried out to look for an infection. In rare occasion, cerebrospinal fluid can be normal despite the presence of an inflammatory CNS disease.

**How can inflammatory CNS disease be treated?**

Treatment of inflammatory CNS disease depends on the primary cause. In case of infectious disease, treatment consists mainly of antibiotic or antifungal medications. Unfortunately we do not have specific treatment for viral infections. In case of non-infectious (pathogen-free inflammatory) disease, treatment is based on trying to counteract the 'over-excitation' of the immune system by giving immuno-suppressing drugs. High dose corticosteroids (prednisone) therapy is the mainstay of treatment.

Other drugs such as azathioprine, cytarabine, mycophenolate, cyclosporine, cyclophosphamide can also be used in combination with corticosteroids. The short-term aim of the treatment is to return the patient to normal using high doses of medication. When this aim is achieved, the medium term aim is to slowly reduce the quantity of drug without resulting in relapse. The long-term aim is ideally to taper the patient off all medication, but more realistically may be to find the lowest dose of medication that will keep him/her free of signs. The main risk of using these additional drugs is to shutdown the bone marrow making the patient more prone to infection.

**Can inflammatory CNS disease be cured?**

In the majority of animals, inflammatory CNS disease can be controlled. This means that the patient can lead a normal life but will need to remain on medication for a variable amount of time, from months to years. Many animals will eventually be tapered off all medications. Unfortunately, a small number of animals with too severe inflammation may not improve despite aggressive treatment. A small number of animals that returned to normal and are taken off medication may experience relapses months later.