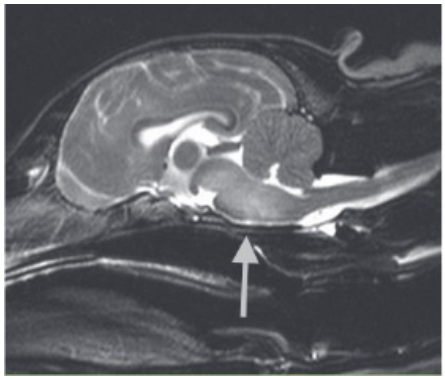
Inflammatory Brain Disease

What is inflammatory brain disease?
Inflammatory brain disease--also known as meningoencephalitis of unknown etiology or "meningitis"-- is an autoimmune disease. An immune system attack on the brain tissue (a.k.a "encephalitis") and the layer surrounding the brain called the meninges (a.k.a "meningitis") causes brain damage and abnormal neurologic signs. Autoimmune inflammation can also occur in the spinal cord (aka "myelitis"), particularly in the neck. Inflammatory brain disease is a broad term that encompasses three specific diseases: granulomatous meningoencephalomyelitis (GME), necrotizing meningoencephalitis (NME), and necrotizing encephalitis (NE). These three different diseases cause slightly different types of brain damage and classically occur in different parts of the brain. Inflammatory brain disease occurs most commonly in small, young to middle aged dogs such as the Maltese, Yorkshire terrier, Chihuahua, Pomeranian, and pug (Pug Dog Encephalitis), but it can occur in larger breed dogs as well.

What are the symptoms of inflammatory brain disease?
The symptoms depend on where in the brain and/or spinal cord the inflammation is located. In dogs with GME, the most common signs are decreased alertness, difficulty maintaining balance, and neck pain. The most common signs in dogs with NME are seizures, behavior changes, and circling.

How is inflammatory brain disease diagnosed?
The only way to definitively diagnose inflammatory brain disease is to collect a sample of brain tissue (i.e. a brain biopsy), but this procedure is rarely performed. MRI of the brain can reveal changes suggestive of inflammation in the brain. Spinal fluid analysis is very useful and often shows high numbers of inflammatory cells, although it can occasionally be normal.


The arrow on this MRI points to inflammation of the brain (white area in the base of the brain).

How is inflammatory brain disease treated?
Treatment involves lifelong suppression of the immune system. High doses of steroids are used initially to try to get the inflammation under control. Response to treatment is demonstrated by improvement or normalization of the neurologic exam and a decrease in the number of cells in the spinal fluid. Additional immunosuppressive medications are added to allow the dose of steroids to be tapered down while maintaining immunosuppression. The advantage to using other medications is that they don't have the negative side effects of prednisone (increased drinking, increased urination, increased appetite, panting, thinning of hair, weight gain). One of the most common immunosuppressive medications is called cytosine arabinoside (Cytosar) which is given as a subcutaneous injection twice daily for two days every 3-4 weeks. Some animals require additional immunosuppressive medications. Inflammatory brain disease is a condition that is controlled rather than cured; therefore discontinuing immunosuppression is strongly discouraged.

What is the prognosis?
The prognosis is variable and depends partially on the variant of the disease. NME and NE generally carry a poorer prognosis than GME. Many patients respond well to treatment and return to normal or nearly normal neurologically. Some patients show no response to therapy and continue to get worse. A third subset of patients shows partial response or initial response but signs progress over time.