Granulomatous Meningoencephalitis and Necrotizing Encephalitis

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BASIC INFORMATION

Description

Granulomatous meningoencephalitis (GME) and necrotizing encephalitis (NE) are disorders that arise from inflammation of the brain and/or spinal cord and their coverings (the meninges). These diseases affect predominantly young, small-breed dogs.

Causes

Both GME and NE are presently considered to be autoimmune disorders, meaning that the body launches an abnormal attack against its own nervous system tissues. Potential triggers for the autoimmune attack may include infections and vaccinations. Genetics may also play a role in the development of GME and NE.

Although any breed of dog may be affected with GME, toy breeds are affected most often. Breeds commonly affected by NE include the pug, Maltese, Yorkshire terrier, Chihuahua, French bulldog, shih tzu, and Pomeranian.

Clinical Signs

Clinical signs are variable and relate to the area of the brain or spinal cord that is inflamed. Clinical signs often worsen rapidly and may include seizures, lethargy, circling, loss of balance, and decreased vision. Both GME and NE can be fatal, with or without aggressive therapy.

Diagnostic Tests

The breed and age of dog and the variety of neurologic signs may create an initial suspicion of GME or NE. A definitive diagnosis cannot be made without evaluation of brain tissue, which requires a biopsy. Because a brain biopsy is not practical in most dogs, information from other tests is valuable for establishing a presumptive diagnosis:

- Routine blood tests are often performed to evaluate the overall health of the affected dog and to eliminate other diseases.
- The key tests to establish a presumptive diagnosis are brain imaging, such as magnetic resonance imaging (MRI) and computed tomography (CT scan), and analysis of spinal fluid (CSF). MRI is the most sensitive method of brain imaging, but a CT scan may also be utilized. They often reveal areas of inflammation. Both GME and NE can affect any region of the brain, but they each have a tendency to involve particular areas, which may help tell them apart.
- Although CSF analysis may rarely be normal, it usually reveals elevated numbers of white blood cells and elevated protein levels. CSF analysis also helps to rule out other diseases (such as tumors and infections) that may affect the nervous system.

TREATMENT AND FOLLOW-UP

Treatment Options

Because GME and NE are caused by inflammation, treatment is initially directed at suppressing the inflammation with steroids. High doses are used initially; as the animal improves, the dosage may be lowered gradually over the course of months. The goal of therapy is to reduce the dosage to the minimal amount needed to control the clinical signs. Side effects of steroids include increased thirst, appetite, and urination. Animals may breathe heavily and pant excessively. Long-term side effects include a thin hair coat, poor wound healing, and muscle loss. Steroid therapy can also make the dog prone to infections of the skin and urinary tract. Notify your veterinarian if any of these side effects occur.

Affected dogs are often treated with additional immunosuppressive medications such as arabinoside-C (*Cytosar*), cyclosporine, or procarbazine. Arabinoside-C and procarbazine are chemotherapeutic drugs, so care must be taken to avoid human exposure to these medications. Dogs with seizures also may require anticonvulsant therapy. (See handout on **Seizures: Treatment**.)

Follow-up Care

Initially, rechecks are scheduled based on the severity of signs and the drugs used. More frequent visits may be necessary for dogs receiving arabinoside-C or procarbazine, because these drugs can have serious side effects, including decreased white blood cell counts, loss of appetite, vomiting, and diarrhea. Careful monitoring with frequent blood tests is indicated. Once the dog responds to therapy and the signs improve, treatment and recheck intervals may be extended.

Prognosis

GME and NE are difficult to cure, but clinical signs often are controllable with aggressive treatment. Treatment may decrease the severity of the clinical signs; however, some neurologic abnormalities may not completely resolve.

The long-term prognosis is guarded to poor, in that most dogs require lifelong treatment and some dogs do not respond even to aggressive therapy. Dogs that do respond to treatment can live for years with these diseases. Relapses may occur after an initial response to treatment, so notify your veterinarian if any clinical signs return. Without treatment, clinical signs usually worsen and can result in death within a few days to weeks.

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