

Dr. Roughie's Questions and Answers

Steroid Responsive Meningitis-Vasculitis: The Disease With Many Names

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Soon after the introduction of the Petit Basset Griffon Vendeen to the United States, owners of the breed reported remarkably similar accounts of painful episodes, noted primarily in young dogs, often associated with fevers, high white blood cells counts and pain most pronounced in the neck region. Many affected dogs experienced recurrent painful episodes while others experienced only a single episode. It is my belief that many of these early cases of juvenile neck pain, often called "the neck thing" early on, were misdiagnosed by the attending veterinarians as Lyme disease, intervertebral disc disease, vague bacterial infections and even trauma. Many of the veterinarians that attended these patients were both unfamiliar with the breed and unfamiliar with similar juvenile pain syndromes seen in other breeds such as the Bernese Mountain Dog and Beagles. Since those early cases of "the neck thing" there has been growing acceptance of the fact that our breed appears to be predisposed to what may best be classified at this point in time as steroid-responsive meningitis, although as we will soon learn, meningitis may not be present in all cases.

Historically, vasculitis (inflammation of blood vessels) affecting the nervous system of dogs was described as far back as 1968. Names that have been applied to nervous system vasculitis of dogs include polyarteritis, systemic necrotizing vasculitis, aseptic meningitis, steroid-responsive meningitis-arteritis, and Beagle pain syndrome. This variation in nomenclature refers to what are very similar diseases. The nomenclature has varied according in great part to how the disease was characterized by the researcher, i.e. whether the description was primarily clinical, i.e. characterized by the clinical signs or response to treatment, versus pathological, i.e. characterized by the lesions noted.

The cause or causes of steroid-responsive meningitis in dogs are poorly understood. It is possible that an infectious agent or other environmental factor may be involved in the development of the disease, although to date, all attempts to isolate viral or bacterial agents have been unsuccessful. It has also been theorized that the disease, or a predisposition to it, may be inherited. These possibilities, which at first appear contradictory, or not exclusive to each other, as it is quite possible that both may be true. In other words, some animals may be genetically predisposed to handle an environmental challenge in a unique way that then leads to disease. What does appear to be likely from more recent studies is that an immune event, specifically directed to the central nervous system occurs in affected animals. Some animals affected with steroid responsive meningitis/vasculitis show lesions of vasculitis elsewhere, to include joints, coronary arteries, skin and testicles. Another finding has been hemorrhage within the spinal canal.

Clinical signs of the disease include fever, neck rigidity and pain, generalized or poorly localized pain and stiff gait or reluctance to move. In more chronic forms of the disease, spinal cord abnormalities, manifested as partial or complete limb paralysis may be noted.

In other breeds, but not to my knowledge in PBGVs, there have been rare reports of cranial nerve deficits. The disease primarily affects dogs between 6 months and 2 years of age, although cases have been reported in other breeds as late as 7 years of age. The disease has a natural waxing and waning course and the severity of episodes is variable between dogs.

Diagnosis of the disease is both challenging and expensive. The expense and risk involved in the thorough investigation of dogs exhibiting recurrent juvenile painful episodes has been the primary factor limiting the knowledge of the disease in our breed. The diagnosis may be made by a combination of history, clinical signs, exclusion of other diseases, and laboratory evaluation of both blood and in severe cases, cerebrospinal fluid. White blood cell counts are often increased in the acute form of the disease. Serum protein electrophoresis may reveal increased alpha₂-globulin levels, which is supportive, but not diagnostic, of the disease. Cerebrospinal fluid analysis classically reveals increased protein levels, increased alpha₂-globulin levels and increased white cell counts. Bacterial culture of the cerebrospinal fluid is negative. In more chronic forms of the disease, diagnosis is more difficult as white blood cell counts both of blood and cerebrospinal fluid may be normal, although cerebrospinal fluid protein levels may remain slightly increased. Of particular value in confirming the diagnosis is the combination of increased alpha₂-globulins in both serum and cerebrospinal fluid.

Dogs affected with aseptic meningitis-vasculitis often respond dramatically to antiinflammatory drug therapy. Nonsteroidal antiinflammatory agents such as carprofen (Rimadyl) or etololac (Etogesic) may be useful for mild cases and in animals experiencing their first episode of pain. After the first relapse, or with the first occurrence where clinical signs are severe, corticosteroid therapy, preferably with prednisone or prednisolone, is indicated. Dosage is tailored to the patient and to the severity of the disease, but typical starting dosages should be immunosuppressive, i.e. at least 4mg/KG per day, gradually decreasing over a period of weeks to months to alternate day therapy. For dogs where meningitis has been confirmed, corticosteroid therapy should ideally not be discontinued for at least six months and after normal cerebrospinal fluid examinations have been confirmed. In the most severe cases, other immunosuppressive agents, such as azathioprine, may be a useful adjunct to corticosteroid therapy.

Although the prognosis for most dogs affected with steroid-responsive meningitis-vasculitis is good, more severely affected dogs may suffer greatly during painful episodes. Owners of dogs with frequent and persistent relapses are often discouraged, both by the side effects of treatment as well as the costs of the diagnostic workups. Rarely, dogs may develop severe complications from the disease to include blindness and rear limb paralysis. Some affected dogs have the potential to bite during painful episodes. Death, either from complications of the disease, or from euthanasia, have been reported.

There are a number of challenges to the study of this disease in Petits Bassets Griffons Vendeens. First, we must define the characteristics and the spectrum of the disease in our

breed, making comparisons to the syndromes seen in other breeds. The relative scarcity of our breed has also been a hindrance to the study of the disease. As previously indicated, making the diagnosis is often cost prohibitive and invasive, particularly if cerebrospinal fluid analysis is necessary. As knowledge of the disease expands in all breeds, there is hope that affordable diagnostic tests may be developed to detect those dogs that may either be subclinically affected or those dogs where more routine laboratory investigation fails to yield a conclusive diagnosis. If indeed it is confirmed that the disease is inherited, determination of the mode of inheritance would be of immediate benefit in the selection of breeding stock. Depending on the mode of inheritance, genetic testing may also offer hope in identifying carrier animals. If environmental factors contribute to the disease, determining which environmental contributors could be avoided and which ones modified would prevent the needless suffering of affected dogs.