Masticatory Muscle Myositis

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Masticatory Muscle Myositis (MMM) is a relatively common inflammatory disorder affecting primarily dogs, rarely cats. This autoimmune disorder targets the muscle of mastication, specifically the temporalis, masseter, and medial and lateral pterygoid muscles. Pets present with common clinical signs at various stages of disease, with the acute phases of disease, in particular, calling for a prompt diagnosis, early intervention, and the best chances for improvement.

Common signalments are younger, larger breed dogs, including the German Shepherd, Labrador Retriever, Golden Retriever, and Doberman Pinscher, although MMM can affect any dog at any age. A possible MMM variant has been reported to affect young (~12 weeks old) Cavalier King Charles Spaniels; MMM is rarely reported in cats. Common historical complaints and clinical signs in the early phases of the disease include lethargy, hyporexia/anorexia, exophthalmos, third-eyelid protrusion, trismus, regional lymphadenomegaly, and dysphagia. Fever is possible. Swelling of the pterygoid muscles might explain the exophthalmos, whereas muscle spasms might explain the reduction in jaw movement seen in trismus. In chronic stages of disease, masticatory muscle atrophy can be severe, with loss of muscle support for the eye leading to enophthalmos and varying limits to open mouth.

Interestingly, there have been anecdotal observations of a relative 'latent' period between the acute and chronic phases, whereby the pet seems partially improved compared to the acute phase.

Differential diagnoses for MMM include inflammatory causes (retrobulbar abscess, generalized polymyositis, extraocular myositis), musculoskeletal (temporomandibular joint disorder), endocrine (hyperadrenocorticism, hypothyroidism), neoplasia (cachexia, trigeminal nerve sheath tumor) and sarcopenia.

The pathogenesis of MMM has been detailed. The muscles of mastication, specifically the temporalis, masseter, and medial and lateral pterygoid muscles, differ in their particular isoforms from their appendicular counterparts. The masticatory muscles originate from the branchial arch mesoderm, whereas those of the appendicular skeletal originate from the paraxial mesoderm. The masticatory muscles have a unique myosin isoform, the Type 2M myofiber, which are subsequently targeted by autoantibodies in MMM, leading to inflammation and phagocytosis, with resultant myonecrosis with end-stage fibrosis. Lymphocytes, particularly T cells, are a key player in MMM. The exact etiology of MMM is unknown, although molecular mimicry between Type 2M myosin inform and an unknown infectious agent has been theorized.

A minimum database should be submitted following a general physical examination that suspects MMM, including a CBC, serum chemistry with CK, urinalysis, and thyroid panel. An inflammatory leukogram may be present, with a chemistry supporting muscle involvement (hyperCKemia, elevated ALT and AST enzyme activity). The serologic test for 2M auto-antibodies is the gold standard for MMM and should be submitted early in the diagnostic workup before starting corticosteroid therapy to minimize false negative results. A temporalis muscle biopsy should be considered early in diagnostic workup, considering the longer turnaround time for serum 2M autoantibody testing and the lack of information pertaining to both severity and prognosis on serum testing alone. Advanced imaging, namely MRI or CT, can be beneficial and sensitive in early stages of disease, helping to identify affected regions of a particular muscle and identifying affected muscle that can be targeted for biopsy to minimize false negative results. Electrophysiology, namely electromyogram (EMG), can be performed with predictable abnormalities that might be identified in the affected muscle.

Response to treatment can be good if instituted early in the disease. Autoimmune therapy is the cornerstone of therapy, with autoimmune corticosteroid dosing commonly instituted until jaw mobility and jaw pain normalize. Slow tapering or corticosteroids over extended months are encouraged to minimize relapse. Adjunctive immunomodulatory drugs, such as leflunomide, mycophenolate, and/or ciclosporin, to name a few, can be beneficial, either replacing the need for corticosteroid altogether or to help lower the dosing of corticosteroid quicker than might be realized is using corticosteroid monotherapy.

Follow-up with these patients' families and follow-up physical examinations are important, paying particular attention to drug side effects, drug monitoring, body weight, degree of muscle atrophy, jaw comfort, and mobility.

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