



HISTIOCYTIC DISEASES

Histiocytic disorders are an emerging group of likely related but confusing cancers reported in dogs. Histiocyte refers to cells of the macrophage/monocyte or “scavenger” cells of the immune system. These cells are present in many tissues of the body, including skin, lymph nodes, lungs, liver, spleen, and bone marrow. Several different “syndromes” have been recognized that may be variations of the same disease or related to the same cell of origin. Histiocytic diseases are currently separated into 5 syndromes: idiopathic periadnexal multinodular granulomatous dermatitis, cutaneous histiocytosis, systemic histiocytosis, splenic histiocytosis/fibrohistiocytic nodules, and malignant histiocytosis.

Idiopathic Periadnexal Multinodular Granulomatous Dermatitis (IPMGD)

IPMGD is a benign disease and may represent an autoimmune disorder. Benign lesions are confined exclusively to cutaneous tissues and arise most commonly on the head. There is no breed or sex predilection. Average age at diagnosis is 6 years. No underlying infectious agents have been identified. Lesions have been reported to regress spontaneously in some cases. Most cases are responsive to corticosteroid therapy. For some patients, continuous, intermittent low dose corticosteroid therapy is required to prevent recurrence of lesions.

Cutaneous Histiocytosis

Cutaneous histiocytosis represents a benign, diffuse infiltration of histiocytes that occur on multiple locations in the skin as nodules and plaques. Cutaneous histiocytosis often occurs in younger dogs, with the Golden retriever and German shepherd breeds overrepresented. Similar to IPMGD, CH is highly responsive to corticosteroid therapy. Long term maintenance therapy may be required to prevent recurrence of lesions.

Systemic Histiocytosis

Systemic histiocytosis is a proliferative disorder of histiocytes that occurs predominantly in middle aged Bernese Mountain Dogs, but is not inclusive to this breed and has been observed in Golden Retrievers, Doberman pinscher, and Rottweilers. Systemic histiocytosis may represent a variant of malignant histiocytosis. SH is a nodular disease confined to the skin, peripheral lymph nodes, and eyes. Skin lesions most commonly involve flank, muzzle, nose, eyelid, and scrotum. SH tends to have a waxing and waning course that is characterized by periods of relative lesion-free remission followed by recrudescence of lesions and symptoms. In addition to skin lesions, symptoms include lethargy, inappetance, and weight loss. Most owners elect euthanasia because of the chronic debilitating nature of the disease. SH is poorly responsive to therapy. Average survivals are 9-10 months.

Splenic Histiocytosis/ Fibrohistiocytic Nodules

Splenic histiocytosis/fibrohistiocytic nodules include a diffuse infiltration of the spleen with histiocytes and a similar, although nodular, form. Golden retrievers, Labrador retrievers, German Shepherds, and cocker spaniels appear to be over-represented. Clinical signs include lethargy, abdominal distension due to enlargement of the spleen, or collapse if the spleen ruptures. Although the spleen can be surgically removed, most dogs will have evidence of metastases to other organs. Average survival is 3-5 months.

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Chemotherapy has not proven to be beneficial in improving survival.

Malignant Histiocytosis

Malignant histiocytosis is a rapidly progressive, widely metastatic proliferation of malignant histiocytes. Malignant histiocytosis is commonly seen in the Bernese Mountain Dog, but other breeds such as the Golden Retriever, Rottweiler, and Doberman pinscher are at increased risk. Symptoms are non specific and include lethargy, inappetance, weight loss, difficulty breathing and neurologic signs. The clinical course is rapid and uniformly fatal. Response to chemotherapy has been unrewarding. Doxil (liposomal encapsulated doxorubicin) has resulted in responses in some patients; however, responses are short-lived and the cost of this drug is prohibitive. Survivals range from a few weeks to several months.

Malignant Fibrous Histiocytosis (MFH)

Malignant fibrous histiocytosis has been described as a soft tissue sarcoma. However, a recent study describes a possible association between malignant histiocytosis and MFH. Comparison of breed and organ involvement was similar suggesting either a similar precursor cell or the possibility that one or both of these diseases can differentiate toward the other disease. MFH has been observed in the retriever breeds (Golden, Labrador, and flat coated) and Rottweiler. In these cases, wide spread metastases typical of MH is common. The majority of dogs do not survive longer than 1 year.

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