Histiocytic Diseases

Histiocytic proliferative disorders (HPDs) are a group of diseases that have a variety of different clinical behaviors. Thus, sometimes it can be confusing for an owner to understand and frustrating for a veterinarian to diagnose and treat.

Histiocytes are immune cells that arise mostly from bone marrow precursor cells and then differentiate into different cell lines. They are present in many different organs including the blood, bone marrow, lymph nodes, spleen and liver. These cells are present all over the body in organs such as the skin, lymph nodes, lungs, liver, spleen, and bone marrow.

Canine cutaneous histiocytoma is usually a benign condition of young dogs that presents as a solitary lesion that eventually regresses on its own. There is a metastatic form but this is rare.

Cutaneous histiocytosis is a benign condition that presents with one or more skin lesions comprised of histiocytes. These occur on one or more locations. This disease often occurs in younger dogs. This disease is usually treated with steroids and other immune suppressive treatments. It can sometimes spontaneously regress and is occasionally treated with surgery as well.

Systemic histiocytosis is a non-cancerous disease in Bernese Mountain Dogs similar to cutaneous histiocytosis, but patients also present with other organs additionally involved. This occurs predominantly in middle aged dogs and also occurs in Golden Retrievers, Rottweilers, Irish Wolfhounds, and, rarely, other breeds. Lesions can be found in most organs. Dogs often present with anorexia, eye inflammation, breathing difficulty, weight loss, lymph node enlargement, weight loss, and lethargy. High calcium levels, anemia, and monocytosis are consistently reported. This is a progressive disease similar to Langerhans histiocytosis in humans. It is treated with immune suppressive therapy, though treatment can be frustrating and often unrewarding. It has a more indolent course than malignant histiocytosis, and the lesions tend to wax and wane.

Malignant fibrous histiocytosis (MFH) is considered a soft tissue sarcoma. MFH has been observed in retrievers and Rottweilers. They often originate in the spleen and metastases are common.
Histiocytic Sarcoma is a malignant disease with different presentations. It may a localized disease with potential metastatic spread or a disseminated disease that affects multiple sites. The disseminated form has also been called malignant histiocytosis.

Histiocytic sarcoma most commonly occurs in Bernese Mountain Dogs, Rottweilers, Flat Coated Retrievers and Golden Retrievers. Dogs are usually middle-aged or older though young dogs have been diagnosed as well. Lesions can occur in most organ but most commonly the joints, spleen, liver, bone marrow, lungs, lymph nodes, central nervous system, and skin. Clinical signs vary according to the organs involved. Generally, patients have lethargy, decreased appetite, weight loss, fever, vomiting, lethargy, and increased lymph nodes. They may also present to a veterinarian with bleeding or severe anemia similar to a patient with autoimmune mediated diseases. Diagnosis is usually achieved by cytology or biopsy. Special stains applied to a patient’s sample may be required to definitively diagnose certain tumors. Complete staging is recommended since this is a tumor that can spread to many organs. Tests include blood work, urinalysis, 3-view chest x-rays or CT scan, abdominal ultrasound, and a bone marrow aspirate if there are indications. Common blood work abnormalities include low platelet counts, increased liver enzymes, low albumin levels, and increased white blood cell counts. Sometimes patients have high calcium levels.

With disseminated disease, the course is rapidly progressive and uniformly fatal. With localized forms, there is the potential for long term control with adequate treatment. However, studies indicate surgery alone is usually inadequate. In one study of joint tumors, treatment with surgery alone gave patients a 6 month survival with 91% chance of metastatic spread. Thus, it makes logical sense to additionally treat with chemotherapy to “mop up” residual disease. As the disease has been better studied, reports indicate an up to 46% response to certain chemotherapy agents.

In cats, these diseases are rare. Most cats present with disseminated forms and aggressive disease.

Malignant fibrous histiocytosis (MFH) is a tumor that contains both histiocytes and fibroblastic cells and is considered to be a soft tissue sarcoma. These are now often called undifferentiated sarcomas or malignant spindle cell tumors. These occur most commonly in the skin, but in the dog can also occur in internal organs such as the spleen. Splenic forms tend to be much more aggressive and metastatic. In cats, these tumors may develop at injection sites. Tumor grade (determined by the pathologist who reads the biopsy) plays an important role. Chemotherapy in addition to local treatment with surgery and/or radiation is often recommended.