Thrombocytopenia, Immune-Mediated

ABOUT THE DIAGNOSIS

Cause: Thrombocytes are commonly called platelets—they are microscopic blood cells in the bloodstream that normally make blood clot, in order to prevent bleeding, such as after a cut or scrape. Thrombocytopenia is an inadequate number of platelets in the blood, which leads to excessive bleeding. It is a problem similar to hemophilia—even minor injury or bruising may lead to significant blood loss because the body is unable to stop the bleeding normally.

In immune-mediated thrombocytopenia, the immune system destroys platelets. Normally, the immune system is responsible for recognizing and destroying foreign invaders such as bacteria and viruses. However, in animals with immune-mediated thrombocytopenia, the immune system is erroneously programmed to work against platelets and the immune system destroys platelets until they are only present in dangerously low numbers. Small blood vessels can then leak blood into surrounding tissues in response to the most minor trauma, leading to significant blood loss from the bloodstream.

In terms of triggers of the disease, there are two types of immune-mediated thrombocytopenia: primary and secondary. Primary immune-mediated thrombocytopenia has no known cause. Secondary immune-mediated thrombocytopenia is triggered by other events such as certain infections, medications, and cancer. The symptoms of either form may range from mild to life-threatening (see Signs to Watch For below).

This disease is diagnosed far more commonly in dogs than cats. It affects female dogs more often than males, and cocker spaniels, poodles, and Old English sheepdogs more often than other breeds although any dog can be affected. Most dogs are middle-aged when diagnosed.

Diagnosis: Immune-mediated thrombocytopenia is suspected based on symptoms of blood loss (excessive bruising; unstoppable bleeding from the gums, nose, or elsewhere; coughing up blood; or more vague signs such as lethargy or decreased appetite). Thrombocytopenia is confirmed on a standard blood test when the platelet count is extremely low. The results of several tests as well as a complete history, thorough physical exam, and the response to treatment are necessary to pinpoint any underlying cause to help arrive at a diagnosis because there are other reasons that could explain a low platelet count, such as disease of the bone marrow (where new blood cells of all types are made). Often, a more advanced degree of testing, involving such tests as bone marrow aspiration and screening for evidence of tick-borne diseases like ehrlichiosis, is necessary to eliminate several "impostor" syndromes that can produce a low-platelet count as part of a totally different disease process. Be sure to share all of your dog's or cat's medical history as well, including all medications given.

Your veterinarian may recommend other tests to evaluate the possibility of various triggers of immune-mediated thrombocytopenia because identifying any of these triggers offers the possibility of eliminating the root cause of the problem, which is the most effective approach.

LIVING WITH THE DIAGNOSIS

Usually, animals will spend a few days in the hospital after receiving this diagnosis, or at least they will be rechecked very frequently until the platelet count comes up to a safe level. Once your pet goes home, give medication exactly as directed by your veterinarian. When a trigger for the problem is not identified (which is fairly common), oral medications given for months or sometimes for years may be required. Many medications must be given in gradually decreasing dosages when the decision is made to discontinue them. Suddenly stopping them can have severe, life-threatening consequences.

Follow your veterinarian's instructions to limit your pet's activity level as appropriate. Although playing and exercise seem like healthy activities, in animals with uncontrolled immune-mediated thrombocytopenia (typically in the first days or weeks of the disease), these activities cause trauma to tiny blood vessels and bleeding.

Most dogs will go into a remission of disease with treatment, but this can be a fatal condition. Dogs can die as a result of losing too much blood, or as a result of bleeding excessively into vital organs such as the brain or spinal cord. Luckily, many dogs permanently recover. For others, this disease may recur weeks to months after recovery. Continue to observe closely for the recurrence of the original symptoms.

TREATMENT

If a specific drug is suspected to be the trigger of immune-mediated thrombocytopenia, it must be discontinued. If an infectious agent is suspected as the root cause, an antibiotic typically is started until test results are received to confirm or eliminate that suspicion. If no specific cause of thrombocytopenia is found, then the diagnosis "by exclusion" is idiopathic thrombocytopenia and the goals of treatment are: to suppress the immune system, reducing platelet destruction; to replace lost blood, if necessary; and to make the patient as comfortable as possible during the initial stages. It is important to protect the affected individual since even very minor trauma (mild bumping of the head or chewing on sticks, for example) can cause severe and/or internal bleeding in the early stages of this disease. If treatment is effective, the platelet numbers return to normal and the risk of bleeding is as low as in any normal pet.

In immune-mediated thrombocytopenia, the problem involves the immune system and its unnecessary destruction of healthy platelets. Therefore, the cornerstone of treatment is reduction and moderation of the overactive immune system. To suppress the immune system, a corticosteroid (e.g., prednisone or dexamethasone) may be given. Other immunosuppressive drugs may be necessary if the side effects of corticosteroids are intolerable or in addition to corticosteroids in cases that are more severe. Blood transfusions can be necessary to replace lost blood if there has been substantial internal or external bleeding. Platelet transfusions are not commonly used in immune-mediated thrombocytopenia because they are quickly destroyed by the disease process.

Other treatments are available. Each patient responds uniquely to medications. Therefore, the combination of treatments will depend on your pet's own response to medications and overall health status. You can consider having a second opinion from a veterinary internal medicine specialist if the cause or treatment remains unclear, or for the latest treatment options. Your veterinarian can refer you to one of these specialists (directory: www.acvim.org or www.vetspecialists.com [North America], www.ecvim-ca.org [Europe]).

DOs

- Realize that this is a life-threatening disease that will require long-term daily treatment for many months, at least. Even if the animal goes into remission and no longer needs medication, there can be disease recurrence.

- If you notice any of the signs listed in Signs to Watch For or if you suspect a relapse, take your pet to your veterinarian or to the local veterinary emergency clinic immediately.
- Inform your veterinarian if your dog or cat has ever been diagnosed with a medical condition and is taking medication. Provide the name(s) of the medication(s).
- Give medication exactly as directed by your veterinarian, and if you are concerned about possible negative effects, discuss them with your veterinarian immediately rather than simply discontinuing the treatment.
- Keep all scheduled recheck appointments; careful monitoring during recovery is vital.
- Be sure to refill prescriptions before you run out of medication. Even a brief period without treatment can cause disease relapse.
- Let your veterinarian know if the side effects of medications (thirst, excessive urination, panting, etc.) are intolerable. Often, other immunosuppressive medications can be added on to treatment that allow the dose of corticosteroids to be reduced.

DON'Ts

- Don't allow your pet, no matter how energetic or normal-looking, to be active until the immune-mediated thrombocytopenia is under control, meaning that a blood test shows that the platelet count is back to a safe range. Premature return to activity can cause internal bleeding.
- Do not postpone getting veterinary treatment if you observe any symptoms of immune-mediated thrombocytopenia. Prompt diagnosis and treatment may prevent complications that are more severe.
- Do not give medication that you have at home that has been prescribed for human use; some of these may interfere with treatment and cause even more severe problems.

WHEN TO CALL YOUR VETERINARIAN

- If you cannot keep a scheduled appointment.
- If you are unable to give medication as directed.

SIGNS TO WATCH FOR

- Watch for general signs of illness, which include vomiting, diarrhea, decreased appetite, weight changes, and changes in behavior such as hiding more than usual and aggressiveness.
- Watch for signs of immune-mediated thrombocytopenia, which include lethargy; weakness; bruises on the gums and skin (petechiae or ecchymoses); blood in stools (including melena—a black, tarry, gelatinous, and metallic-smelling type of stool that indicates bleeding in the stomach or intestine), vomitus (hematemesis), or urine (hematuria); or any other abnormal bleeding, respiratory difficulty, and exercise intolerance.

ROUTINE FOLLOW-UP

- Follow-up appointments are scheduled to monitor progress and to determine if treatment should be adjusted. Abnormalities on previous tests can be pursued at this time, and the exact timing and interval is determined individually. Usually, rechecks will be very frequent initially (even every few days), but as the pet improves these will be scheduled further apart.

ADDITIONAL INFORMATION

- When immune-mediated thrombocytopenia occurs at the same time as immune-mediated hemolytic anemia (caused by the immune system attacking red blood cells), the condition is called Evans syndrome.
- Most animals recover from immune-mediated thrombocytopenia, but up to 20% of these patients will die. The prognosis is worse if Evans syndrome is present (both immune-mediated thrombocytopenia and immune-mediated hemolytic anemia).

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