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Mastocytosis

Mastocytosis is the term for a group of disorders caused by too many "mast cells." It is most common in children, but the more important forms are seen in both children and adults. Mast cells are found throughout the body, but are especially plentiful in the skin and digestive tract. Mast cells are part of the immune defense system. They are also found around wounds and seem to play a role in healing of injuries.

Mast cells work by releasing chemicals that attract white blood cells to areas of the body where they are needed. Researchers also think mast cells may have a role in the growth of blood vessels. No one with too few or no mast cells has ever been found, which indicates to some scientists that having too few mast cells may be incompatible with life.

The presence of too many mast cells, or mastocytosis, can occur in two forms: cutaneous and systemic. The most common cutaneous (skin) form is also called urticaria pigmentosa, which occurs when mast cells infiltrate the skin. Systemic mastocytosis is caused by the accumulation of mast cells in the tissues and can affect organs such as the liver, spleen, bone marrow, and small intestine.

Urticaria pigmentosa was first described in 1869, while systemic mastocytosis was first reported in scientific literature in 1933. The true incidence of either type of mastocytosis remains unknown but mastocytosis generally is considered to be an "orphan disease" (Orphan diseases affect approximately 200,000 or fewer people in the United States).

Symptoms

Chemicals released by mast cells because physiological changes that leads to typical allergic responses such as hives, itching, abdominal cramping, and even shock. When too many mast cells exist in a person's body, the additional chemicals can cause bone pain, abdominal discomfort, nausea and vomiting, ulcers, diarrhea, skin lesions, and episodes of hypotension (very low blood pressure and faintness) or anaphylaxis (shock).

Diagnosis

Urticaria pigmentosa can be diagnosed through the abnormally high concentration of mast cells in the skin. The diagnosis of systemic mastocytosis can be made by a biopsy showing an increased number of mast cells in an organ other than the skin. The biochemical hallmark of the disease is an elevation of histamine or mast cell tryptase protein in blood. Urine may also contain high levels of histamine metabolites (products) as well as metabolites of prostaglandin D2. Elevations of plasma histamine or mast cell protease are occasionally seen in patients with allergic diseases, but, unlike mastocytosis patients, their plasma histamine levels are elevated only temporarily. Plasma tryptase and histamine levels are persistently increased in mastocytosis patients. Other tests that are important in the evaluation of a suspected case of mastocytosis include a bone marrow examination and a bone scan. Special stains, such as Giemsa and toluidine blue, are used on a bone marrow sample to demonstrate the increase in marrow mast cells that occurs in a large percentage of people with the disease.

Treatment

An array of drugs is used to treat the symptoms of mastocytosis, including antihistamines (to prevent mast cell release of chemicals), and anticholinergics (to relieve intestinal cramping). A number of drugs are used for the treatment of specific symptoms of mastocytosis.

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- Itching and skin complaints are frequently treated with drugs called HI antihistamines such as hydroxyzine.
- Ulcer-like symptoms are relieved by another type of antihistamine called H2 anti-ulcer antihistamines such as cimetidine or ranitidine.
- Severe flushing and hypotension (low blood pressure) are treated prophylactically (before symptoms appear) with H1 and H2 antihistamines and with epinephrine after symptoms begin.
- Skin lesions that are cosmetically disturbing to the patient can be temporarily reduced through use of topical and sometimes intravenous steroids. If the patient has only a few lesions, a doctor may remove them surgically.
- Malabsorption, or impaired ability to take in nutrients, can be treated with steroids. In rare cases in which mastocytosis is malignant, or cancerous, prednisone and/or chemotherapy is necessary.