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Commentary

Aggressive Systemic Mastocytosis: Types, Symptoms, Diagnosis and Treatment

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DESCRIPTION

Aggressive Systemic Mastocytosis (ASM) may be a clonal mastocyte disease characterized by progressive growth of neoplastic cells in diverse organs resulting in organopathy. The organ-systems most often affected are the bone marrow, skeleton, liver, spleen, and therefore the alimentary canal.

Systemic mastocytosis (mas-to-sy-TOE-sis) may be a rare disorder that leads to too many mast cells build up in your body. A mastocyte may be a sort of white blood corpuscle. Mast cells are found in connective tissues throughout your body. Mast cells help to guard you from disease and help your system function properly and normally.

When you have systemic mastocytosis, excess mast cells build up in your skin, bone marrow, alimentary canal or other body organs. When triggered, these mast cells release substances which will cause signs and symptoms almost like those of an allergy and, sometimes, severe inflammation which will end in organ damage. Common triggers include alcohol, spicy foods, insect stings and certain medications.

Signs and symptoms of systemic mastocytosis depend upon the a part of the body suffering from excessive mast cells. Too many mast cells can build up within the skin, liver, spleen, bone marrow or intestines. Less commonly, other organs like the brain, heart or lungs also could also be affected.

TYPES OF SYSTEMIC MASTOCYTOSIS

The five main sorts of systemic mastocytosis include.

Indolent systemic mastocytosis

This is often the foremost common type and typically doesn't include organ dysfunction. Skin symptoms are common, but other organs could also be affected, and therefore the disease may worsen slowly over time.

Smoldering systemic mastocytosis

This sort is related to more-significant symptoms and should include organ dysfunction and worsening disease over time.

Systemic mastocytosis

With another blood or bone marrow disorder this severe type develops rapidly and is usually related to organ dysfunction and damage.

Aggressive systemic mastocytosis

This rare type is more severe, with significant symptoms, and is typically related to progressive organ dysfunction and damage.

Mast cell leukemia

This is often a particularly rare and aggressive sort of systemic mastocytosis.

Signs and symptoms of systemic mastocytosis may include:

- Flushing, itching or hives
- Abdominal pain, diarrhea, nausea or vomiting
- Anemia or bleeding disorders
- Bone and muscle pain
- Enlarged liver, spleen or lymph nodes
- Depression, mood changes or problems concentrating

The mast cells are triggered to supply substances that cause inflammation and symptoms. People have different triggers, but the foremost common ones include:

- Alcohol
- Skin irritation
- Spicy foods
- Exercise
- Insect stings
- Certain medications

CAUSES OF SYSTEMIC MASTOCYTOSIS

Most cases of systemic mastocytosis are caused by a random change

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(mutation) within the KIT gene. Typically this flaw within the KIT gene isn't inherited. Too many mast cells are produced and build up in tissues and body organs, releasing substances like histamine, leukotrienes and cytokines that cause inflammation and symptoms.

Complications of systemic mastocytosis can include:

- Anaphylactic reaction
- Blood disorders
- Peptic ulcer disease
- Reduced bone density
- Organ failure

Diagnosis include:

To diagnose systemic mastocytosis, your doctor will likely start by reviewing your symptoms and discussing your medical record, including medications you've taken. He or she will then order tests that search for high levels of mast cells or the substances they release. Evaluation of organs suffering from the condition also could also be done.

Tests may include:

- Blood or urine tests
- Bone marrow biopsy
- Skin biopsy
- Imaging tests like an X-ray, ultrasound, bone scan and CT scan
- Biopsy of organs suffering from the disease, like the liver
- Genetic testing

Treatment of systemic mastocytosis include:

Treatment may vary, counting on the sort of systemic mastocytosis and therefore the body organs affected. Treatment generally includes controlling symptoms, treating the disease and regular monitoring.

- Controlling triggers
- Medications
- Chemotherapy
- Stem cell transplant
- Regular monitoring