

UNDERSTANDING MASTOCYTOSIS

Mastocytosis is a Rare Immune Disorder and Affects Males and Females in Equal Numbers

Mastocytosis is Considered to be an Orphan Disease, Affecting 200,000 or Fewer People in the United States

Mastocytosis is an uncommon accumulation of mast cells in the skin and other parts of the body that can cause itching, pain and other symptoms. Learn more below about this rare condition, symptoms and treatment.

01 | Types and Causes

There are two main types of Mastocytosis:

- **Cutaneous** – this form is more commonly found in children and affects the skin. Mast cells collect in areas of the skin, forming bumps (urticarial pigmentosa).
- **Systemic** – this form usually occurs in adults when mast cells accumulate in the skin, stomach, intestine, liver, spleen, lymph nodes or bone marrow. Bone marrow disruption can lead to serious blood disorders, including leukemia.

02 | Symptoms and Diagnosis

The symptoms of Cutaneous Mastocytosis are restricted to the skin and range from mild skin rash to more severe symptoms including blistering, hypotension, gastrointestinal bleeding and anaphylactic shock.

In cases of Systemic Mastocytosis, patients may present with a number of symptoms including enlarged liver or spleen, organ function impairment and musculoskeletal pain.

Diagnosis is dependent on what the indicators are present and may include the following:

- Skin and bone marrow biopsies
- Measurement of mast cell mediators in blood & urine
- Blood count

- Liver function studies
- Genetic tests

03 | Treatment

Cutaneous Mastocytosis is usually treated with antihistamines and resolves with no other treatment necessary. There is no cure Systemic Mastocytosis; however, the disease can be controlled with H₁ and H₂ blockers to relieve itching and reduce gastric acid production. In aggressive cases, interferon-alpha may be injected under the skin to reduce effects on bone marrow.

For more information, please visit:

<http://www.aaaai.org/conditions-and-treatments/related-conditions/systemic-mastocytosis>

Did You Know?
Systemic Mastocytosis was first reported in 1949

References

<https://rarediseases.org/rare-diseases/mastocytosis/>

<http://www.merckmanuals.com/home/immune-disorders/allergic-reactions-and-other-hypersensitivity-disorders/mastocytosis>

<https://my.clevelandclinic.org/health/articles/mastocytosis>