

[Home](#) > [Types of Cancer](#) > [Mastocytosis](#) > Mastocytosis - Introduction

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Mastocytosis - Introduction [1]

This section has been reviewed and approved by the [Cancer.Net Editorial Board](#) [2], 06/2016

ON THIS PAGE: You will find some basic information about this disorder, which may or may not become cancerous, and the parts of the body it may affect. This is the first page of Cancer.Net's Guide to Mastocytosis. To see other pages, use the menu. Think of that menu as a roadmap to this full guide.

Mast cells are a type of blood cell made in the bone marrow. They are involved in allergic reactions and help fight off parasitic infections. Mast cells produce histamine, a chemical that can cause itching, sneezing, congestion, swelling, and wheezing.

About mastocytosis

Mastocytosis describes a group of disorders that are caused by too many mast cells in the body. The number of mast cells in the body can increase as a result of some non-cancerous (benign) or reactive conditions. However, certain types of cancers begin when healthy mast cells change and grow out of control.

There are 2 general types of mastocytosis:

- **Cutaneous mastocytosis.** This type of mastocytosis involves the skin.
- **Systemic mastocytosis.** This type of mastocytosis involves the entire body.

Cutaneous mastocytosis

Cutaneous mastocytosis is an increase of mast cells in the skin. About 90% of people with mastocytosis have the cutaneous type. Subtypes of cutaneous mastocytosis include:

- **Urticaria pigmentosa.** This is the most common type of cutaneous mastocytosis. The main sign of urticaria pigmentosa is the appearance of tan or red-brown spots on the skin. These spots usually develop on the midsection of the body first and then spread over the rest of the body. A person with this type of mastocytosis may also experience nausea, vomiting, and diarrhea.
- **Solitary mastocytoma.** This type of mastocytosis is more common in infants and children than in adults. It usually causes a large nodule, about 3 to 4 centimeters (cm) in diameter, to form on an arm or leg. This nodule is called a mastocytoma.
- **Diffuse erythrodermic mastocytosis.** This type of mastocytosis is found most often in children younger than 3 years old. Diffuse erythrodermic mastocytosis may not be noticeable when a child is born, but it can later show up as a rapid thickening of the skin. Symptoms of systemic mastocytosis (see the [Symptoms and Signs](#) [3] section) and blisters are also common.
- **Telangiectasia macularis eruptiva perstans.** This type of cutaneous mastocytosis develops mainly in adults. The most common sign is lesions that do not itch and are smaller than those of urticaria pigmentosa.

Systemic mastocytosis

Systemic mastocytosis involves internal organs throughout the body, including the gastrointestinal tract, bone marrow, liver, spleen, and lymph nodes. Lymph nodes are tiny, bean-shaped organs that help fight infection. About 85% of the time, urticaria pigmentosa (see above) develops first.

Depending on the number of mast cells in an organ, the disease is classified as either indolent, which means it is slow growing, or aggressive, which means it is fast growing. As the number of mast cells builds up in an organ, the symptoms of the disease may get worse.

Systemic mastocytosis can become cancerous. The risk of systemic mastocytosis becoming cancerous is 7% when the disease begins in childhood and as much as 30% in adults. Mast cell leukemia involves the blood, while mast cell sarcoma involves the body's soft tissues.

The [next section in this guide is Statistics](#) [4]. It helps explain how many people are diagnosed

with this condition. Or, use the menu to choose another section to continue reading this guide.

Links

[1] <http://www.cancer.net/cancer-types/mastocytosis/introduction>

[2] <http://www.cancer.net/about-us>

[3] <http://www.cancer.net/node/19227>

[4] <http://www.cancer.net/node/19225>