

# MYCOSIS FUNGOIDES

## What Is Mycosis Fungoides?

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma (CTCL). Although the terms MF and CTCL are often used interchangeably, this can be a source of confusion. CTCL is an umbrella term. All patients with MF have CTCL, but not all patients with CTCL have MF.

MF follows a slow, chronic (indolent) course and very often does not spread beyond the skin. About 10% of the time, MF can progress to lymph nodes and internal organs. Symptoms of MF can include flat, red, scaly patches, thicker raised lesions called plaques, and sometimes large nodules or tumors. MF can look like other common skin conditions like eczema or psoriasis, and might be present for years or even decades before it's diagnosed as CTCL. The disease can progress over many years, often decades.

There is continuing research into possible environmental or infectious agents that might contribute to MF, but at this time no single factor has been proven to cause this disease. One theory about how MF might occur is because of chronic low-level stimulation of T-cells in the skin. The skin has about twice the number of T-cells than blood - about a million per cm<sup>2</sup>. These T-cells are very busy, responding to outside insults and protecting our largest organ from the outside world. Sometimes some of the T-cells get stimulated - for example by an insect bite, or topical allergen - and that can cause chronic low level stimulation. There is no supportive research indicating that MF is hereditary. Exposure to Agent Orange may be a risk factor for developing MF for veterans of the Vietnam War, but no direct cause-effect relationship has been established.

It is important to know that MF is not contagious. It is not an infection and cannot be passed from person to person.

## How Common Is Mycosis Fungoides?

CTCL is a rare family of diseases. While the number of new patients with CTCL diagnosed each year is relatively low (about 3,000), it is estimated that, since patients have a very long survival, there may be as many as 30,000 patients living with cutaneous lymphoma in the United States and Canada.

MF is more common in men compared to women, in black people compared to white people, and in

patients older than 50 years of age compared to younger people.

Due to the difficulty of diagnosing the disease in its early stages, the slow course of MF, and the lack of an accurate reporting system, these numbers are probably low estimates.

## What Does Mycosis Fungoides Look Like?

One of the challenging things about MF is that it doesn't look the same for all patients. MF can appear anywhere on the body, but tends to affect areas of the skin protected from sun by clothing. Patches, plaques and tumors are the clinical names for different skin manifestations and are generally defined as "lesions."

- **Patches** are usually flat, can be smooth or scaly, and look like a "rash."
- **Plaques** are thicker, raised, usually scaly lesions. MF patches and plaques are often mistaken for eczema, psoriasis or "non-specific" dermatitis until an exact diagnosis is made.
- **Tumors** are raised "bumps" or "nodules" which may or may not ulcerate (form sores).

While it is possible to have all three types of lesions at the same time, most people who have had the disease for many years experience only one or two types of lesions, generally patches and plaques. Only rarely are tumors the first lesion.

A common symptom of MF is itching, with 80 percent or more of people with mycosis fungoides suffering from itching.

## How Is Mycosis Fungoides Diagnosed?

Mycosis fungoides is very difficult to diagnose, especially in early stages. The symptoms and skin biopsy findings of MF are similar to other benign skin conditions like eczema, psoriasis, parapsoriasis, or pityriasis lichenoides. Patients may go for years or even decades before a definitive diagnosis of MF is established. MF is sometimes diagnosed initially only by dermatologists or oncologists who specialize in cutaneous lymphomas.

Typical procedures done to diagnose MF include:

- A complete physical exam (including a thorough skin exam);
- A skin and/or lymph node biopsy (removal of a small piece of tissue) for examination under the



microscope by a pathologist (a doctor who studies tissues and cells to identify diseases);

- Blood tests - the most common blood tests are a complete blood count (CBC) and a comprehensive metabolic panel (CMP);
- And possibly imaging tests such as CT (computerized axial tomography) and/or PET (positron emission tomography) scans.

It is very important that any diagnosis of MF is confirmed by a pathologist (dermatopathologist or hematopathologist) who has expertise in diagnosing cutaneous lymphomas.

### What Is the Prognosis for Mycosis Fungoides?

For most people, MF is an indolent, chronic disease, but the course of MF for any one individual can be unpredictable. It can be slow, rapid or static. MF almost never progresses to lymph nodes and internal organs without showing obvious signs of progression in the skin such as worsening ulcers or tumors, and examining the skin is a very important clue to possible internal disease.

There is no known cure for MF. Some patients have long-term remission with treatment, and many more live with minimal or no symptoms for many, many years. Research indicates that most patients diagnosed with MF live with early stage disease, and have a normal life span. With advances in research and new treatment options resulting from physician collaboration and clinical trials, MF patients are experiencing better care and an array of effective treatment options that work for them.

### How Is Mycosis Fungoides Treated?

There is no single treatment approach that is optimal for all patients with MF, and treatment choice is individualized.

For effective long-term disease management, choosing the best treatment requires considering the disease manifestations in the skin, blood and lymph node involvement by the disease, the potential adverse effects of the treatment and your other medical conditions (comorbidities), as well as your preferences.

You may be prescribed a single treatment or a combination of treatments.

Selecting the right treatment or combination of treatments for you can be daunting, and it's important to work with a specialist who has experience treating patients with MF.

The treatment of MF is continuously evolving and improving.

At any one time, there are numerous clinical trials ongoing to determine newer and better treatments. It's important to discuss clinical trials with your care provider.

Organizations like the National Comprehensive Cancer Network (NCCN) and the European Organisation for Research and Treatment of Cancer (EORTC) are dedicated to improving the care provided to patients with cancer. These institutions, and also the British Association of Dermatologists (BAD), work to provide and maintain up-to-date guidelines for the care of patients with MF.

### Skin-Directed Therapies

In most patients with MF, the lymphoma cells are primarily limited to the skin, and one can have excellent and long-lasting responses with treatments directed at the skin, or "skin-directed therapy." Because treatment is directed just at the skin, the toxicity of these treatments is low. Examples of skin-directed therapies are creams, ointments, or gels that are applied to the skin, such as topical steroids, topical nitrogen mustard, retinoids, and immune stimulating creams (imiquimod). Ultraviolet light (phototherapy) and radiation therapy are also types of skin-directed therapy.

### Systemic Therapies

"Systemic therapy" refers to any treatment that, after absorption, reaches the bloodstream and is therefore distributed across the body "system." Any drug that is taken by mouth, given as a suppository, injected under the skin, taken under the tongue, or directly infused through a blood vessel will eventually reach all body organs and tissues (including the skin), with the exception of the brain, which by design is protected by a specific "barrier." Systemic therapies are used in MF when skin-directed therapies aren't working well enough, are too difficult to apply, or the disease is advanced. Systemic therapies may be used alone or in combination, and are often used together with skin-directed therapies (for example, using a pill to make you more sensitive to ultraviolet light therapy).

There are many examples of systemic therapies that are used in MF, including extracorporeal photopheresis, targeted/biologic agents given by infusions (histone deacetylases and antibodies), oral medications (retinoids, and methotrexate), and subcutaneous injections (interferon). Traditional or standard chemotherapy, which would be used for other blood cancers, is only rarely used in MF because of severe side effects and a high rate of MF returning after treatment.

### Reviewed and updated:

July 2024 - *Cutaneous Lymphoma Foundation's Writers Bureau*



**CUTANEOUS LYMPHOMA FOUNDATION**

PO Box 969 • Warren, MI 48090

Email: [info@clfoundation.org](mailto:info@clfoundation.org)

Phone: 248.644.9014

Website: [www.clfoundation.org](http://www.clfoundation.org)