

FOLLICULOTROPIC MYCOSIS FUNGOIDES

What is Folliculotropic Mycosis Fungoides?

Cutaneous lymphomas primarily arise in the skin. Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma (CTCL). It is an indolent (slow) type of CTCL, and generally follows a chronic course. Folliculotropic mycosis fungoides (FMF) is a subtype of MF that involves hair follicles.

Who Gets Folliculotropic Mycosis Fungoides? How Common is it?

Cutaneous lymphomas are a relatively rare compared to other cancers, with only 1500-2000 new cases diagnosed each year across North America. The median age of presentation is over 50 years old, with a higher occurrence in men compared to women. Since the disease has a slow course, there are probably many more people living with the disease. It is estimated that 30,000 people in the United States and 3,000 people in Canada are living with a diagnosis of cutaneous lymphoma. There is general consensus that these numbers are low estimates, because they do not account for misdiagnosed or undiagnosed patients. It is difficult to diagnose FMF in its early stages, and there is not an accurate reporting system. About ten percent of MF patients have FMF.

There is no clear known cause for MF or FMF. Cutaneous lymphomas are not contagious and cannot be passed from one person to another, and they generally are not inherited.

What Does Folliculotropic Mycosis Fungoides Look Like?

MF lesions include flat, red, scaly patches, thicker raised lesions (plaques), and sometimes larger nodules or tumors. Patients with FMF might also notice areas of hair loss, especially around the face or scalp, pimples or blackheads, or increased infections within their plaques because of involvement of the hair follicles. Patients with FMF might experience itching. Patients may also experience no specific symptoms related to their skin rash.

How is Folliculotropic Mycosis Fungoides Diagnosed and Staged?

Many of the same procedures used to diagnose and stage other types of CTCLs are used in FMF, including a physical exam and history; 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); and a series of imaging tests such as CT (computerized axial tomography and/or PET (positron emission tomography) scans to determine if the cancer has spread to lymph nodes or other organs. When examined under the microscope, skin biopsies from FMF patients show involvement around hair follicles (“folliculotropism”).

It is important to confirm the diagnosis of FMF by a dermatopathologist or a hematopathologist who has expertise in diagnosing lymphomas.

The following staging system is used to determine the extent of FMF:

Stage IA—Less than ten percent of the skin is covered in red patches and/or plaques.

Stage IB—Ten percent or more of the skin surface is covered in patches and/or plaques.

Stage IIA—Any amount of the skin surface is covered with patches and/or plaques; lymph nodes are enlarged, but the cancer has not spread to them.

Stage IIB—One or more tumors are found on the skin; lymph nodes may be enlarged, but the cancer has not spread to them.

Stage III—Nearly all of the skin is reddened and may have patches, plaques or tumors; lymph nodes may be enlarged, but the cancer has not spread to them.

Stage IVA—Most of the skin area is reddened and there is involvement of the blood with malignant cells or any amount of the skin surface is covered with patches, plaques or tumors; cancer has spread to the lymph nodes and the lymph nodes may be enlarged.

Stage IVB—Most of the skin is reddened or any amount of the skin surface is covered with patches, plaques or tumors; cancer has spread to other organs; and lymph nodes may be enlarged whether cancer has spread to them or not.



What Causes Folliculotropic Mycosis Fungoides?

Although there is continuing research, currently no single factor is known to cause cutaneous lymphomas. They are acquired diseases with no clear genetic or hereditary link, and they are not contagious. Despite a number of anecdotal reports, there does not seem to be a clear or defined link between cutaneous lymphoma and chemical exposure, pesticides, radiation, allergies, the environment, or occupations. Exposure to Agent Orange may be a risk factor for developing MF, but no direct causal relationship has been shown.

How is Folliculotropic Mycosis Fungoides Treated?

FMF is generally treated similar to other forms of MF, but need to be monitored more closely because of involvement of hair follicles. Most patients should first be treated with skin directed treatments like light (photo)therapy and topical agents. If skin directed treatments don't work, then systemic agents (pills, injections, or intravenous treatments) are added. The list of systemic agents for CTCLs is growing, and includes retinoids, interferon injections, histone deacetylase inhibitors (vorinostat and romidepsin), and antibody-based therapies, among others.

Skin radiation is also commonly used to treat FMF. Skin radiation for FMF is usually a low dose method (electron beam) that spares the internal organs.

A very small number of patients with FMF need more aggressive therapy with chemotherapy or a stem cell transplant to control their disease.

What is the Prognosis of Folliculotropic Mycosis Fungoides?

The prognosis of FMF is determined by the type and the extent of skin involvement and overall stage. Early stage FMF has a prognosis very similar to classic MF, with excellent survival. People with more advanced FMF (thicker lesions or tumors or involvement deeper in the skin) are more likely to progress to more advanced stages. Large cell transformation is a sign of progression, and may indicate more serious disease. Involvement of organs other than the skin (such as the lungs or liver), lymph nodes, or blood also indicate more serious disease.

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs and finding the best treatments for patients with rare diseases like cutaneous lymphoma. Physicians may recommend clinical trials because standard treatments may not be effective.

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*August 2018 - Cutaneous Lymphoma Foundation's
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