

## Mycosis Fungoides (Alibert-Bazin Syndrome, Granuloma Fungoides) — Signs and Symptoms

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**Skin lesions have a broad range of etiology and can be complex in diagnosis due to their similar appearance on inspection. Cutaneous T-Cell lymphoma is a rare skin disease that presents with several distinct manifestations. Most common of this manifestation is Mycosis Fungoides and Sezary syndrome. Mycosis fungoides is a slow progressing set of mushroom-shaped skin lesions, while Sezary syndrome is a rapidly progressing generalized skin disease with a worse prognosis.**



### Introduction to Mycosis Fungoides

#### Skin lesions in your USMLE test

USMLE skin lesions will present on your test with visual pictures or in-depth descriptions of them. Usually, the **questions will cover the pathophysiology and presentation** or can include the immunologic or cytologic features of the lesion. Most of the lesions will be common but, occasionally, rare lesions will pop up as a question.

A rare but potential group of skin lesions revolve around Cutaneous T-cell lymphoma (CTCL) which can present as Mycosis Fungoides (MF) or Sezary syndrome. **CTCL is characterized as a form of non-Hodgkin's lymphoma** where neoplastic T-lymphocytes begin to localize to the skin. There are a few other forms of CTCL with provisional classifications but, for the purpose of this resource and preparation for the USMLE, this article will focus only on MF and Sezary syndrome.

## General Overview of Mycosis Fungoides

**High Yield:** CTCL is most commonly CD4+ (Helper) T cell-based in 65% of cases. It is more common in patients with sub-Saharan ancestry and there is a 2:1 predominance towards men than woman.

CTCL is **extremely rare in children and most commonly occurs in middle aged to elderly patients**. Due to the nature of the skin lesions, the average time before a diagnosis is made is 6 years. Prognosis is poor since there is no cure for either MF or Sezary syndrome and terminal outcomes depended upon type and stage of the disease. Differentials to keep in mind include allergic and irritant contact dermatitis, lichen planus, and plaque psoriasis.

## Definition of Mycosis Fungoides

MF is an indolent form of non-Hodgkin's lymphoma that presents with skin lesions in a stepwise fashion. MF is the **most common presentation of CTCL and comprises 44% of all cases**. The name originates from the presentation of the patches of plaques that were first thought to resemble a mushroom shape.

**Note:** There is also some confusion where CTCL is called MF and it should be clear that MF is a sub-form of CTCL. MF has an incidence of 0.36 per 100,000 cases in the USA. The three stepwise presentations occur in the following pattern:

1. **Patch presentation** with dermatitis that is located to the trunk and often times on the buttocks, which are usually not pruritic.
2. **Plaque presentation** that is characterized by intense pruritus and can be associated w/ lymphadenopathy.
3. **The Tumor phase** is the last manifestation that results in lesion changes to the point of ulceration.

## Diagnosis of Mycosis Fungoides

Diagnosis can be difficult since the initial presentation can be similar to so many other skin related diseases. Due to the indolent nature, it is important to take a thorough history to understand the progression of lesions. A biopsy of the lesion will show distinct characteristics. **Diagnostic testing would include a CBC, LFTs, flow cytometry, and HIV screening**. In suspected later stages of the disease, chest X-ray and potential CT scans can be done to look for further organ involvement.

## Management of Mycosis Fungoides

Management of MF depends upon the stage:

- **Stage 1** management focuses on topical therapy with antipruritic medications

and retinoids.

- **Stage 2 and 3** include the use of steroids, surgical excision, and radiotherapy for advanced disease.

There is some research that has shown allogeneic stem cell transplants have helped with MF treatment. Photochemotherapy with UV-A (PUVA) has been beneficial for some patients.

## Prognosis of Mycosis Fungoides

Prognosis is generally good due to the indolent and slower nature of the disease. Stages determine the prognosis with early stages being better than later.

**USMLE quick review:** Due to the sometimes complex nature of skin lesions, it is important to remember the common ones that will be tested on the USMLE. It can be helpful to review their visual and microscopic appearance to help reinforce your understanding of them!

### Review - common skin lesions:

- **Macule:** A flat lesion that has clear circumscribed margins that differ from surrounding skin color and is less than 1cm in size.
- **Patch:** Larger sized macules that are greater than 1cm and well-circumscribed (seen in MF).
- **Papule:** An elevated skin lesion less than 1cm in size. Commonly thought of as Basal cell carcinoma.
- **Plaque:** A larger papule greater than 1cm in size.
- **Vesicle:** A fluid-filled blister that is less than 1cm in size.
- **Bulla:** A larger fluid-filled blister greater than 1 cm in size.
- **Pustule:** A pus containing vesicle that can be of any size.
- **Wheal:** Smooth papule with a transient time course usually occurs secondary to an allergic reaction and seen in cases of hives.
- **Scale:** Flakey skin lesion that is dry and usually seen in eczema and in squamous cell carcinoma.
- **Crust:** Exudate from an underlying skin infection that crusts over.

## Sezary Syndrome as a Special Form of Mycosis Fungoides

**Sezary syndrome is an aggressive form of CTCL** that will present with rapid symptoms of generalized edematous skin, lymphadenopathy, generalized alopecia, and hyperkeratosis of palms and plantar surface of the foot. In some patients, it can present with ectropion of the eyelids and hepatosplenomegaly. It comprises 5% of all CTCL presentations.

### Diagnosis

**Diagnosis is made by analyzing the Sezary cell count** (cerebriform specific cellular morphology). Immune cell phenotypic expression changes with increased CD4 to CD8 ratio and a loss of CD markers 2, 3, 4, and 5. In advance disease, a chest X-ray and CT may be indicated for further spread of the disease.

## Management

Management focuses on how fast the disease is progressing. Therapy involves immune modulation with IFN- $\alpha$  and antineoplastic drugs that include chlorambucil, methotrexate, and etoposide. Topical immunomodulators can be used, such as imiquimod and retinoid agents.

## Prognosis

Prognosis is poor due to the incurable nature of the disease. Since Sezary syndrome is rapidly progressive and results in immunosuppression, patients will often die due to secondary systemic infections from Staph aureus and Pseudomonas. **The median survival from the date of diagnosis is 2–4 years.**

## References

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[Sezary Syndrome](#) via radiopaedia.org

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