

Mycosis Fungoides / Sézary Syndrome

Mycosis fungoides is a cutaneous T-cell lymphoma, which is characterized by infiltration of CD4+ cells in the skin. It typically affects patients between the ages of 40 to 60, and patients develop pruritic skin lesions, rashes, plaques and patches. Biopsy may reveal Pautrier's abscesses. This may progress to a T-cell leukemia, known as Sézary syndrome. A diagnostic finding for Sézary syndrome are Sézary cells with "cerebriform" nuclei seen in a peripheral blood smear.



PLAY PICMONIC

Mechanism

Neoplastic CD4+ Cells

[Abnormal CD \(4\) Fork](#)

This is a disorder of unusual expression of CD4+ T cells. Mycosis fungoides and Sézary syndrome are subtypes of cutaneous T-cell lymphomas, distinguished through severity and leukemic involvement. Sézary syndrome has a worse prognosis than mycosis fungoides.

Adults 40 - 60 Years Old

[40 to 60 Display](#)

Mycosis fungoides and Sézary syndrome have an average age of onset between 40 and 60 years of age. It is extremely rare for the disease to appear before the age of 20. Often with Sézary syndrome, patients may present in their 60's, having already developed mycosis fungoides earlier in their lives.

Mycosis Fungoides

Neoplastic CD4+ Cells Infiltrate Skin

[Abnormal CD \(4\) Fork in Skin](#)

In mycosis fungoides, neoplastic CD4+ T-cells infiltrate the skin. Diagnosis is difficult in early phases of disease, as the rashes, plaques and patches resemble eczema or psoriasis.

Rashes Or Plaques

[Rashes and Plaques](#)

Patients develop pruritic plaques, patches, and discolored lesions with mycosis fungoides. Early phases of disease often resemble eczema or psoriasis.

Pautrier's Abscesses

[Party Abscesses](#)

Pautrier's abscesses, or microabscesses, are visible under a microscope, and are described as well-defined collections of mononuclear mycosis cells located within the epidermis. These findings are useful in diagnosing mycosis fungoides.

Sézary Syndrome

Neoplastic CD4+ Spread from Skin to Systemic Involvement

Abnormal CD (4) Fork in Blood

With infiltration into the bloodstream, mycosis fungoides and Sezary syndrome prognosis can worsen as the tumor burden increases. Sezary disease is defined by the presence of CD4+ T-cell infiltrates in the skin, an absolute Sezary cell count of 1000 cells/mm or higher, CD4 cells with a CD4/CD8 ratio of 10 or higher and expanded CD4 T-cell abnormality on peripheral blood smear.

Sézary Cells With "Cerebriform Nuclei"

Scissor Cell with Cerebral Nucleus

On peripheral blood smear, a diagnostic finding in patients with Sezary syndrome are Sezary cells, which are described as having a prominent nuclear cleft. This clefting has lead to the term "cerebriform nuclei" as a description.