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 Sezary syndrome. A diagnostic finding for Sezary syndrome are Sezary cells with "cerebriform" nuclei seen in a peripheral blood smear.



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Mechanism

Neoplastic CD4+ Cells
Abnormal CD (4) Fork
This is a disorder of unusual expression of CD4+ T cells. Mycosis fungoides and Sezary syndrome are subtypes of cutaneous T-cell lymphomas, distinguished through severity and leukemic involvement. Sezary syndrome has a worse prognosis than mycosis fungoides.

Adults 40 - 60 Years Old
40 to 60 Display
Mycosis fungoides and Sezary 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 Sezary 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.

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