

Mycosis fungoides, a CTCL subtype, may progress in $\frac{1}{3}$ of patients

within the skin, or even beyond skin to other parts of the body^{1,2}

MF and SS are rare lymphomas that arise in the skin but may involve other disease compartments¹⁻³
Higher skin stage and multicompartamental involvement are associated with poor prognosis^{1,3}

Progression in MF and Sézary syndrome: Be attentive to these signs and symptoms

Changes in skin lesions or symptoms

- Increase in body surface area with skin lesions³
- Appearance of a new type of lesion⁴
- Changes in the type of lesion (patches, plaques, tumors)³
- Changes in the pigmentation of lesions³
- Reappearance of lesions after remission⁴
 - Patients who are in remission may relapse with a mixture of lesion types
- Onset or worsening erythroderma³
- New or worsening pruritus⁵
 - More common in late-stage MF and SS
 - Not all patients experience pruritus
 - May be an indicator of progression, relapse, or superinfection
- New or worsening burning pain, or sharp “pins and needles” sensation in the skin⁶

New or increased blood tumor burden

- Appearance or increase in detectable levels of Sézary cells in peripheral blood, as determined by flow cytometry³
 - Blood tumor burden may be detectable in all stages of MF; low level blood involvement (B1) may be present in early stage (IA-IIIB) MF⁷
 - High blood burden (B2) in SS or advanced MF (stage IV) is associated with shortened survival¹
- Increase in absolute Sézary cell counts in the peripheral blood, as determined by flow cytometry³
 - Quantification is recommended for any suspected extracutaneous disease³
- T-cell clones in peripheral blood, and presence of identical circulating T-cell receptor clones in the skin and blood, detectable by molecular analysis³
 - Absolute Sézary cell counts and molecular analyses should be interpreted in the context of overall clinical presentation^{3,4,8}

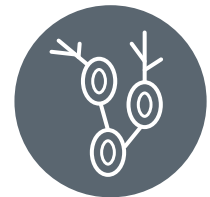
Signs of extracutaneous disease

- Enlarged regional lymph nodes or organomegaly³
 - May be indicative of lymph node or visceral involvement, but should be evaluated in the context of overall clinical presentation³
- Presence, or increased levels, of Sézary cells in the peripheral blood³
 - Blood tumor burden may be detectable in early MF (patch/plaque), or tumor stage⁷

Identifying MF or SS progression involves converging evidence from multiple types of tests. NCCN guidelines recommend a multi-disciplinary approach for diagnosing and managing MF and SS^{3,8}

CTCL=cutaneous T-cell lymphoma;
MF=mycosis fungoides;
SS=Sézary syndrome

Be vigilant for signs of progression in your patients with MF and SS



References: 1. Agar NS, Wedgeworth E, Crichton S, et al. Survival outcomes and prognostic factors in mycosis fungoides/Sézary syndrome: validation of the revised International Society for Cutaneous Lymphomas/European Organisation for Research and Treatment of Cancer staging proposal. *J Clin Oncol.* 2010;28(31):4730-4739. 2. Amorim GM, Niemeyer-Corbellini JP, Quintella DC, et al. Clinical and epidemiological profile of patients with early stage mycosis fungoides. *An Bras Dermatol.* 2018;93(5):546-542. 3. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Primary Cutaneous Lymphomas. V2.2021. 4. Cerroni L. Mycosis fungoides—clinical and histopathologic features, differential diagnosis, and treatment. *Semin Cutan Med Surg.* 2018;37(1):2-10. 5. Serrano L, Martinez-Escala ME, Zhou XA, Guitart J. Pruritus in cutaneous T-cell lymphoma and its management. *Dermatol Clin.* 2018;36(3):245-258. 6. Field H, Gao L, Motwani P, Wong HK. Pruritus reduction with systemic anti-lymphoma treatments in patients with cutaneous T cell lymphoma: A narrative review. *Dermatol Ther (Heidelb).* 2016;6(4):579-595. 7. Scarisbrick JJ, Hodak E, Bagot M, et al. Blood classification and blood response criteria in mycosis fungoides and Sézary syndrome using flow cytometry: recommendations from the EORTC cutaneous lymphoma task force. *Eur J Cancer.* 2018;93:47-56. 8. Larocca C, Kupper T. Mycosis fungoides and Sézary syndrome: an update. *Hematol Oncol Clin North Am.* 2019;33:103-120.