Your patients with adult-onset mastocytosis in the skin may have systemic mastocytosis[1,2]

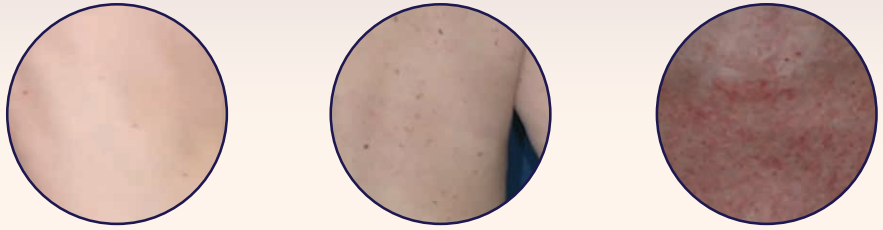
Systemic mastocytosis (SM) is a clonal mast cell disorder characterized by the uncontrolled proliferation and activation of abnormal mast cells.[3,4] ISM represents the largest subtype of the disease.[5-7] SM can present with a range of potentially debilitating symptoms that may affect multiple organ systems—from skin lesions, anaphylaxis, and diarrhea to organ damage.[3,4]

Skin lesions typical of mastocytosis can exhibit swelling, reddening, or pruritus, which can occur when mast cells are degranulated, such as by stroking skin lesions (Darier's sign) or changes in temperature.[3,8-10]

In adult patients with SM who have skin lesions, the lesions are typically monomorphic maculopapular lesions (known as MPCM or UP) and present as small, round, and pigmented.[8,10] The number of lesions may vary among patients, ranging from a few to possibly widespread coverage.[8,10]

Other subvariants include[8,11]:

- DCM
- Mastocytoma



Various skin lesion presentations associated with SM[2]

~90% of patients presenting with adult-onset mastocytosis in the skin may have SM[1,2*†]

*Based on a study from the European Competence Network on Mastocytosis Registry examining 1145 patients with mastocytosis in the skin who underwent a bone marrow biopsy, of whom 944 (82%) were diagnosed with SM.[1]

†Based on a study of 59 patients with the clinical diagnosis of adult-onset mastocytosis in the skin established between 2004 and 2008 at a single center in Germany. 57 of 59 patients (97%) were diagnosed with SM based on WHO criteria.[2]

DCM=diffuse cutaneous mastocytosis; ISM=indolent systemic mastocytosis; MPCM=maculopapular cutaneous mastocytosis; UP=urticaria pigmentosa.

This information is intended for US healthcare professionals only. It does not provide medical advice and is for educational purposes only.

SUSPECTSM

Diagnosis of SM requires the presence of 1 major criterion and at least 1 minor criterion, or at least 3 minor criteria[11‡]

Major criterion	Multifocal dense infiltrates of mast cells (≥15 mast cells in aggregates) detected in sections of bone marrow or other extracutaneous organ(s)
Minor criteria	• >25% of all mast cells are atypical on bone marrow smears or are spindle-shaped in dense and diffuse mast cell infiltrates in sections of bone marrow or other extracutaneous organ(s) • KIT D816 or other activating KIT mutation in bone marrow or other extracutaneous organ(s) • Mast cells in bone marrow, blood, or another extracutaneous organ aberrantly express 1 or more of CD2, CD25, and/or CD30 • Baseline serum tryptase concentration >20 ng/mL in the absence of a myeloid AHN

‡WHO Classification of Tumours Editorial Board. Haematolymphoid tumours [Internet; beta version ahead of print]. Lyon (France): International Agency for Research on Cancer; 2022 [cited September 20, 2023]. (*WHO Classification of Tumours Series*. 5th ed.; vol. 11). Available from: https://tumourclassification.iarc.who.int/chapters/63.

AHN=associated hematological neoplasm; KIT=KIT proto-oncogene, receptor tyrosine kinase.

Skin findings typical of patients with adult-onset cutaneous mastocytosis can be similar to those found in patients with SM, including ISM.[2,8] A bone marrow biopsy and clinical evaluation are necessary to confirm a diagnosis of SM.[11‡]

References: 1. Fuchs D et al. *J Allergy Clin Immunol Pract*. 2021;9(4):1705-1712.e4. **2.** Berezowska S et al. *Mod Pathol*. 2014;27(1):19-29. **3.** Pardanani A. *Am J Hematol*. 2021;96(4):508-525. **4.** Theoharides TC et al. *N Engl J Med*. 2015;373(2):163-172. **5.** Ungerstedt J et al. *Cancers*. 2022;14(16):3942. **6.** Sperr WR et al. *Lancet Haematol*. 2019;6(12):e638-e649. **7.** Cohen SS et al. *Br J Haematol*. 2014;166(4):521-528. **8.** Hartmann K et al. *J Allergy Clin Immunol*. 2016;137(1):35-45. **9.** Gülen T et al. *J Intern Med*. 2016;279(3):211-228. **10.** Akin C, ed. *Mastocytosis: A Comprehensive Guide*. Springer; 2020. **11.** WHO Classification of Tumours Editorial Board. Haematolymphoid tumours. [Internet; beta version ahead of print]. Lyon (France): International Agency for Research on Cancer; 2022 [cited September 20, 2023]. (*WHO Classification of Tumours Series*. 5th ed.; vol. 11). Available from: https://tumourclassification.iarc.who.int/chapters/63

To learn more about identifying and diagnosing SM, please visit SuspectSM.com