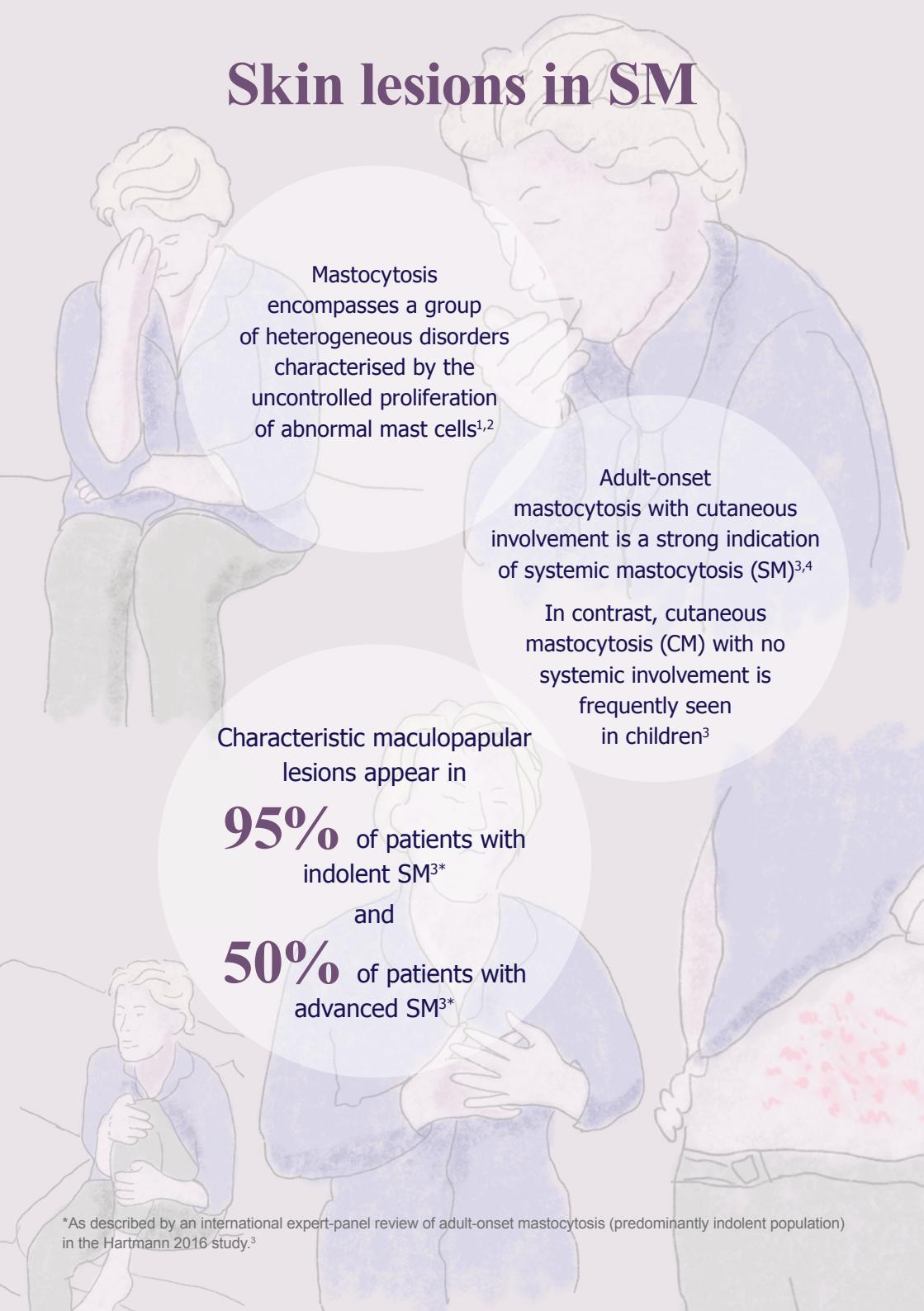




SKIN LESIONS: GETTING TO THE BOTTOM OF UNEXPLAINED SYMPTOMS

Could your patients have SM?

Skin lesions in SM



Mastocytosis encompasses a group of heterogeneous disorders characterised by the uncontrolled proliferation of abnormal mast cells^{1,2}

Adult-onset mastocytosis with cutaneous involvement is a strong indication of systemic mastocytosis (SM)^{3,4}

In contrast, cutaneous mastocytosis (CM) with no systemic involvement is frequently seen in children³

Characteristic maculopapular lesions appear in

95% of patients with indolent SM^{3*}

and

50% of patients with advanced SM^{3*}

*As described by an international expert-panel review of adult-onset mastocytosis (predominantly indolent population) in the Hartmann 2016 study.³

Could you be missing SM?

You may be the first to see these symptoms in initial screening that might trigger suspicion of SM



Image credit: CID, ISM/SCIENCE PHOTO LIBRARY.



Reproduced with permission from ©DermNet www.dermnetnz.org 2023.

MACULOPAPULAR SKIN LESIONS (OR URTICARIA PIGMENTOSA)³

- **Small, round, brown monomorphic lesions** often start to develop on the **thigh**
- **Lesions** may spread to the **trunk and extremities** over the years

Use this information to help you recognise skin lesions and suspect SM^{3*}

Characteristic

Adult-onset mastocytosis

Typical course of disease

Chronic

Typical tryptase level (ng/mL)

>20

Most frequent type of cutaneous lesions

Maculopapular

Typical morphology of maculopapular lesions

Monomorphic

Typical size of maculopapular lesions

Small (1–10 mm)⁵

Typical distribution of maculopapular lesions

Thigh, trunk

*Please note that this table is based on the Hartmann 2016 study on consensus report of the European Competence Network on Mastocytosis, the American Academy of Allergy, Asthma & Immunology and the European Academy of Allergology and Clinical Immunology³; this may not be representative of all SM patients, and the clinical presentation and severity of skin lesions may vary between individuals.

A detailed bone marrow examination is recommended to distinguish between adult CM and SM, and to establish or exclude the diagnosis of SM⁶

Could you be missing SM?

The diagnosis of SM can be easily missed owing to the wide differential diagnosis of skin lesions



Reproduced with permission from ©DermNet www.dermnetnz.org 2023.

DARIER'S SIGN^{3,7}

- Darier's sign, which is manifested as a **wheal-and-flare reaction upon mechanical stroking and rubbing of lesions**, is a highly specific clinical sign of mastocytosis

OTHER SKIN SYMPTOMS⁸

47% of SM patients experience **itching***

44% of SM patients experience **flushing***

42% of SM patients experience **skin changes***

Heat and burning of flushing which indicates onset of a larger mast cell activation attack was a common concern

*Based on data from 149 patients with self-reported SM in the US Mast Cell Connect registry in the Jennings 2018 study.⁸

Recognising initial symptoms is key to achieving early diagnosis of SM

If you recognise persistent unexplained skin lesions, consider ordering a tryptase test or referring the patient to a haematologist for a bone marrow examination⁶

References:

1. Gülen T, et al. *J Intern Med*. 2016;279(3):211–228.
2. Pardanani A. *Am J Hematol*. 2021;96(4):508–525.
3. Hartmann K, et al. *J Allergy Clin Immunol*. 2016;137(1):35–45.
4. Berezowska S, et al. *Mod Pathol*. 2014;27(1):19–29.
5. Orphanet. Maculopapular cutaneous mastocytosis. Available at: https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=GB&Expert=79457. Accessed May 2023.
6. Valent P, et al. *J Allergy Clin Immunol Pract*. 2022;10(8):1999–2012.e6.
7. Galen BT, Rose MG. *Blood*. 2014;123(8):1127.
8. Jennings SV, et al. *Immunol Allergy Clin North Am*. 2018;38(3):505–525.