

## Skin lesions in SM

Mastocytosis encompasses a group of heterogeneous disorders characterised by the uncontrolled proliferation of abnormal mast cells<sup>1,2</sup>

> Adult-onset mastocytosis with cutaneous involvement is a strong indication of systemic mastocytosis (SM)<sup>3,4</sup>

In contrast, cutaneous mastocytosis (CM) with no systemic involvement is frequently seen in children<sup>3</sup>

Characteristic maculopapular lesions appear in

95% of patients with indolent SM3\*

50% of patients with advanced SM3\*

and

<sup>\*</sup>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



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## MACULOPAPULAR SKIN LESIONS (OR URTICARIA PIGMENTOSA)<sup>3</sup>

- 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<sup>3\*</sup>

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 lesi	ions Monomorphic
Typical size of maculopapular lesions	Small (1–10 mm) <sup>5</sup>
Typical distribution of maculopapular lesi	ions Thigh, trunk

A detailed bone marrow examination is recommended to distinguish between adult CM and SM, and to establish or exclude the diagnosis of SM<sup>6</sup>

<sup>\*</sup>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 Immunology<sup>3</sup>; this may not be representative of all SM patients, and the clinical presentation and severity of skin lesions may vary between individuals.

### Could you be missing SM?

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



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#### DARIER'S SIGN<sup>3,7</sup>

 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 SYMPTOMS<sup>8</sup>

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

 $44^{0}/_{0}$  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

<sup>\*</sup>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 examination<sup>6</sup>

#### References:

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