



Mastocytosis, MCAS, and Related Disorders – Diagnosis, Classification and Therapy

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Message from the Guest Editors

Mastocytosis is a heterogeneous group of myelodysplastic diseases with abnormal clonal MC proliferation, activation and accumulation of MC in the skin, bone marrow and/or other visceral organs. The diagnosis is based on the WHO criteria, in which the tryptase level, histopathological and immunophenotypic (CD2/CD25) assessment of MCs and detection of D816V somatic mutation of *KIT* gene are crucial. The disease is divided into 7 variants: cutaneous mastocytosis, indolent systemic mastocytosis (ISM), SM with an associated clonal haematological non-MC-lineage disease (SM-AHNMD), aggressive SM (ASM), MC leukaemia (MCL), MC sarcoma (MCS), and extracutaneous mastocytoma.

Symptoms of MCAS and SM can be managed by blockade of mediator receptors (H1 and H2 antihistamines), leukotriene receptor blockage, inhibition of mediator synthesis or release, anti-IgE therapy, or a combination of these medications. Acute episodes of MC activation require epinephrine. Patients with SM or primary MCAS may need a cytoreductive therapy to prevent severe symptoms including anaphylaxis.

In this special Issue, we will publish research papers, up-to-date review articles, and commentaries are all welcome.





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Message from the Editor-in-Chief

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