Use this tool to record your patients' diagnostic lab results[1]

This tool includes values that are relevant to assessing a diagnosis of SM utilizing WHO criteria, and does not represent medical advice. Please note: these values may or may not be sufficient for an SM diagnosis, and healthcare providers should make all decisions in accordance with their judgment and patient context.

Circle or write in the values that apply

Serum Total Tryptase

_____ ng/mL

Biopsy Location

_____

KIT D816V

Mutation present + / – VAF % _____

High-sensitivity KIT D816V assays are useful when screening for SM.[1]

Mast Cell Burden

Mast cell aggregates + / – % of atypical mast cells _____

Determining mast cell burden is important in suspected cases of SM. CD117 and tryptase are mast cell markers for IHC.[1]

	CD25	CD2	CD30
IHC	+ / –	+ / –	+ / –
Flow cytometry	+ / –	+ / –	+ / –

Atypical expressions of CD25, CD2, and CD30 can indicate the neoplastic nature of mast cells.[1]

IHC=immunohistochemistry; SM=systemic mastocytosis; VAF=variant allele fraction; WHO=World Health Organization.

World Health Organization diagnostic criteria for SM[1]

Diagnosis of SM requires the presence of 1 major criterion and at least 1 minor criterion or at least 3 minor criteria

Major criterion	• Multifocal dense infiltrates of mast cells (≥15 mast cells in aggregates) detected in sections of bone marrow and/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 D816V or other activating KIT mutation in peripheral blood, 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, CD30 • Baseline serum tryptase concentration >20 ng/mL in the absence of a myeloid AHN

It is important to explore the minor diagnostic criteria, as up to approximately **45% of indolent systemic mastocytosis (ISM) cases** may not fulfill the major criterion[1,2]

Subtyping of SM depends on factors such as mast cell burden, organ damage, and signs of myeloproliferation or myelodysplasia. ISM can be determined when there is no organ damage and no associated hematologic malignancy.[3-5]

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

References: 1. WHO Classification of Tumours Editorial Board. Haematolymphoid tumours [Internet]. Lyon (France): International Agency for Research on Cancer; 2024 [cited April 24, 2024]. (*WHO Classification of Tumours Series*, 5th ed.; vol. 11). Available from: https://tumourclassification.iarc.who.int/chapters/63 **2.** Ungerstedt J et al. *Cancers*. 2022;14(16):3942. **3.** Pardanani A. *Am J Hematol*. 2023;98(7):1097-1116. **4.** Valent P. *HemaSphere*. 2021;5(11):e646. **5.** Khoury JD et al. *Leukemia*. 2022;36(7):1703-1719.