Supplemental Table S1: Current classification of primary mastocytosis

<table>
<tr><th colspan="4">Mastocytosis</th></tr>
<tr><th colspan="2">Systemic mast cell activation disease (MCAD)</th><th>Cutaneous mastocytoses (CM)</th><th>Mast cell sarcoma</th></tr>
<tr><th>Systemic Mastocytosis (SM)</th><th>Mast Cell Activation Syndrome (MCAS)</th><td rowspan="2">• maculopapular mastocytosis = urticaria pigmentosa (UP)
• diffuse CM
• solitary cutaneous mastocytoma
• telangiectasia macularis eruptiva perstans</td><td rowspan="2"></td></tr>
<tr><td>• indolent SM
• well-differentiated indolent SM
• smoldering SM
• aggressive SM
• SM with an associated leukemia
• mast cell leukemia</td><td>• with hypertryptasemia
• without hypertryptas-emia</td></tr>
</table>

Based on:

Vysniauskaite M et al. Determination of plasma heparin level improves identification of systemic mast cell activation disease. PLoS One 2015;10:e0124912.
Afrin LB et al. Often seen, rarely recognized: mast cell activation disease--a guide to diagnosis and therapeutic options. Ann Med 2016;48190-201.
Arber A et al. The 2016 revision to the World Health Organization (WHO) classification of myeloid neoplasms and acute leukemia. Blood 2016;127:2391-405.