Systemic Mastocytosis Fact Sheet

What is Systemic Mastocytosis?
- Systemic mastocytosis (SM) is a group of rare diseases in which uncontrolled growth and accumulation of mast cells (a type of white blood cell) occurs in one or more organs. In patients with SM, mast cells can accumulate in multiple organ systems, including the skin, GI tract, spleen, lymph nodes and bone marrow:
  - SM can be characterized into indolent (ISM) and advanced forms. ISM is the benign form of SM and is generally associated with a good prognosis, while advanced SM can be fatal within a few years.
  - The mast cells release substances such as histamine, which can lead to symptoms including itching, fever, abdominal pain, nausea and vomiting.

A Closer Look at Advanced SM
- In advanced SM, mast cells accumulate in such high quantities that they lead to organ damage and dysfunction, bone fractures and anemia. Subtypes of advanced SM include aggressive systemic mastocytosis (ASM), mast cell leukemia (MCL) and SM with an associated hematologic neoplasm (SM-AHN).
- Treatment is tailored towards each individual patient and their disease.
- The major goal of treatment is to control mast cell growth and expansion.

Types of Mastocytosis
- CUTANEOUS: Symptoms affect skin - no organ involvement
- SYSTEMIC: Includes isolated bone marrow mastocytosis (BMM) and smoldering SM (SSM)
- INDOLENT: Includes isolated bone marrow mastocytosis (BMM) and smoldering SM (SSM)
- ADVANCED: ASM, SM-AHN, MCL

Life expectancy not dependent upon disease
- MEDIAN SURVIVAL TIME
  - ISM: 16.5 years
  - ASM: 3.5 years
  - MCL: 2 years
  - SM-AHN: <6 months

Percentage of SM Cases by Subtype
- MCL: 1%
- ASM: 5-10%
- SM-AHN: 20-30%
- ISM: 60-70%

References: