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**
  - Symptom affect skin - no organ involvement
- **SYSTEMIC**
  - Includes isolated bone marrow mastocytosis (BMM) and smoldering SM (SSM)
  - Life expectancy not dependent upon disease
- **INDOLENT**
  - Includes isolated bone marrow mastocytosis (BMM) and smoldering SM (SSM)
- **ADVANCED**
  - ASM
  - SM-AHN
  - MCL
  - Life expectancy dependent upon disease

**MEDIAN SURVIVAL TIME**

- ISM: 16.5 years
- ASM: 3.5 years
- SM-AHN: 2 years
- MCL: <6 months
- MEDIAN SURVIVAL TIME

References: