Systemic mastocytosis (SM) is a rare disease characterized by severe and unpredictable symptoms across multiple systems.1,2

Given the heterogeneity and fluctuations in severity of symptoms, patients with SM may interact with many different clinical specialties over several years before being diagnosed with the disease.3

SM is broadly categorized into non-advanced SM and advanced SM. Most patients (90% to 95%) are considered to have non-advanced SM which primarily includes indolent systemic mastocytosis (ISM) (90% of patients with SM) and a small subset of patients who have asymptomatic SM (ASM).4,5

The objective of this study was to describe the presenting signs, symptoms, and clinical characteristics of patients receiving a diagnosis from a community oncologist within the US Oncology Network.

Methods

Study Design

This was a retrospective observational cohort study of patients with SM treated in The US Oncology Network.

Patients with a diagnosis of SM between January 2010 – May 2021 were eligible for inclusion in the study.

Patients with SM or ISM were categorized as non-advanced SM. Remaining patients were included in the advanced SM cohort.

Eligible patients were followed longitudinally after their SM diagnosis until August 31, 2021, last patient record, or date of death, whichever occurs first.

Date Source

Data were abstracted through chart review from iKnowMed, the electronic health records (EHR) database of The US Oncology Network.

The Limited Access Death Master File of the Social Security Administration was an additional source of vital status (death).

Inclusion/Exclusion Criteria

• Age ≥ 18 years at SM diagnosis

• Not receiving treatment for other documented, SM-unrelated primary health conditions.

• SM diagnosis from a community oncologist within the US Oncology Network.

• Diagnosis of SM

Inclusion/Exclusion Criteria

• Data were abstracted through chart review from iKnowMed, the electronic Data Source

• Eligible patients included in the study.

• Patients with SM or ISM were categorized as non-advanced SM.

• Remaining patients were included in the advanced SM cohort.

• Patients in the non-advanced cohort reported an average of 3.9 symptoms as presentation, while advanced patients presented with 2.6 symptoms on average.

• Advanced SM patients, symptoms and low blood count were listed as a reason for referral in 43.8% of patients. 32% of patients listed elevated blood counts as a reason for referral (Figure 4).

• Among non-advanced patients, 47% of patients were referred due to symptoms and 33.3% of patients were referred due to mastocytosis.

• The symptomology profile differed between the SM cohorts but across both cohorts, reported symptoms were widespread and diverse.

• Among patients with advanced SM, 58.8% reported at least one systemic symptom, 43.6% reported at least one skin symptom, and 31.3% of patients reported at least one neurological or GI symptom at initial presentation.

• Among the non-advanced patients included in this study, 70.6% of patients reported at least one neurological symptom, 64.7% at least one skin symptom, and 47.1% reported at least one GI symptom.

• Patients in the non-advanced cohort reported, an average of 5.9 symptoms as presentation, while advanced patients presented with 2.6 symptoms on average.

• This analysis provides insight into the complex diagnostic journey for SM patients that involves multiple physician specialties.

• Advanced and non-advanced SM patients presented with multiple symptoms across a number of domains. Increased awareness of the multiple specialties involved and the heterogeneous nature of ISM symptoms may have improved the diagnostic journey for patients.