Poster #160

Real-world chart pull data on the clinical presentation and diagnosis of indolent systemic mastocytosis

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SCAN ME

Background/Objectives

- Systemic mastocytosis (SM) is a clonal mast cell neoplasm driven by *KIT* D816V mutation in ~95% of adults with SM.[1] Indolent systemic mastocytosis (ISM) is a mast cell disease characterized by debilitating neurologic, musculoskeletal, gastrointestinal, and cutaneous symptoms and impaired quality of life (QoL).[1,2]
- Due to heterogeneity of symptomology and limited disease awareness, the time between initial symptom onset and receiving a diagnosis of SM has been reported by patients to be approximately 7-9 years.[3]
- The objective of this study was to assess and describe the natural history 'journey' in a real-world clinical practice setting

Methods

- The Integra Connect platform provides oncology practices with a technology platform to help interpret complex datasets and improve value-based patient outcomes. The Integra Connect network currently includes 12 community practices on the East and West coast of the United States.
- IntegraConnect PrecisionIQ invited practicing hematologists in its network to complete a structured electronic case report form (eCRF) for patients that the oncologist treated who were diagnosed with ISM within the past 36 months. The case report captured retrospective data on patients with ISM in a variety of categories (Table 1).
- The eCRF included questions on practice demographics, satisfaction with current treatment options, patient demographics, referral patterns, patient lab values, symptoms, diagnostic process, and treatment regimen.

Table 1. eCRF categories and example questions

Category	Example Questions (Note: not all questions included in the eCRF are shown here.)
Practice-related questions	• How many patients with ISM have you managed in the past 3 years? • What is your primary practice setting?
Diagnosis-related questions	• What laboratory/pathologic/imaging studies were used to make a diagnosis of ISM? • Was the serum tryptase level persistently ≥11.5 ng/mL other than at diagnosis? • What was the patient's KIT D816V mutation status? • Following bone marrow biopsy, what was the percentage of infiltration by mast cells? • Was a biopsy reviewed where the presence of ISM diagnostic criteria were not initially observed, but identified upon consultation/second review? • Was your patient initially diagnosed with ISM?
Symptom-related questions	• Did the patient exhibit any of the following gastrointestinal/musculoskeletal/neuropsychiatric/systemic signs/symptoms at presentation/first visit to you? • Since the initiation of first-line therapy, how have the patient's SM symptoms changed?
Treatment-related questions	• What treatments (class and specific medication) did you prescribe to this patient? • What was the duration of this therapy? • How was the patient's response to therapy measured?

Results

Physician Demographics

Table 2. Physician and Practice Characteristics

	Physicians (n=17)	
	n	%
Primary Practice Setting		
Office-based private practice	11	65%
Hospital-based practice	5	29%
NCCN institution	1	6%
Volume of ISM Patients Managed in Past 3 Years		
1-5	5	29%
6-10	10	59%
11-15	2	12%
Average Number of Other Patients Treated in Past 3 Years		
Cutaneous mastocytosis (CM)	4.8 patients	
Mast cell activation syndrome (MCAS)	2.8 patients	
Myelofibrosis (MF)	15.6 patients	

- The majority of participating oncologists were in office-based practices affiliated with a group purchasing organization (Table 2).
- Most respondents managed between 6-10 patients with ISM within the past 3 year (Table 2).
- All physicians managed at least 1 with MF, but not all providers managed patients with MCAS or CM (Table 2).
- Patients with ISM saw an average of at least 2.2 physician specialties prior to their first visit at the completing oncologist's practice.
- Patients were most commonly referred to a participating oncologist for assessment and treatment by allergists/immunologists and dermatologists (Figure 1).

Results (Continued)

Patient Demographics

- Data were collected on 65 ISM patients identified by participating physicians.
- Most patients were between the ages of 40 and 69 years (Table 3).
- Patients had an average of 2.5 comorbidities. The most common comorbidities (Figure 2) were anxiety (40%), cardiovascular conditions (29%), diabetes mellitus (26%), depression (23%), and chronic kidney disease (22%). 11% of patients had no comorbidities.

Figure 1. Referral Patterns

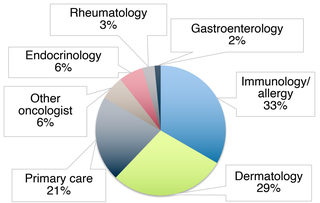
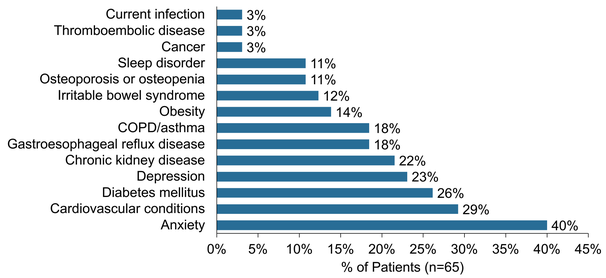


Table 3. Patient Demographics

	ISM Patients (n=65)	
	n	%
Gender		
Male	31	48%
Female	32	49%
Unknown	2	3%
Age		
<40 years	4	6%
40-49 years	15	23%
50-59 years	17	26%
60-69 years	19	29%
70-79 years	8	12%
80-89 years	2	3%

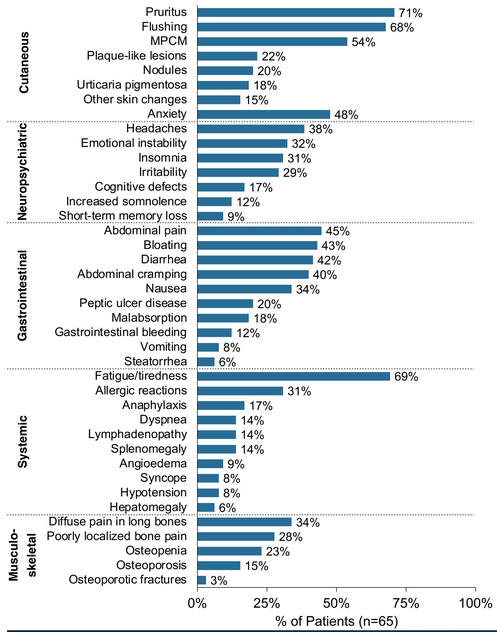
Figure 2. Patient Comorbidities (n=65)



- The most common cutaneous signs/symptoms at diagnosis were pruritus (71%), flushing (68%), and maculopapular cutaneous mastocytosis (MPCM) (54%), and the most common neuropsychiatric signs/symptoms were anxiety (48%) and headaches (38%) (Figure 3).
- The most common gastrointestinal signs/symptoms at diagnosis were abdominal pain (45%), bloating (43%), diarrhea (42%), and abdominal cramping (40%), and the most common musculoskeletal signs/symptoms were diffuse pain in long bones (34%), poorly localized bone pain (28%), and osteopenia (23%) (Figure 3).
- The most common systemic signs/symptoms were fatigue/tiredness (69%) and allergic reactions (31%) (Figure 3).
- Anaphylaxis occurred in 17% of patients (Figure 3).

Signs and Symptoms at ISM Diagnosis

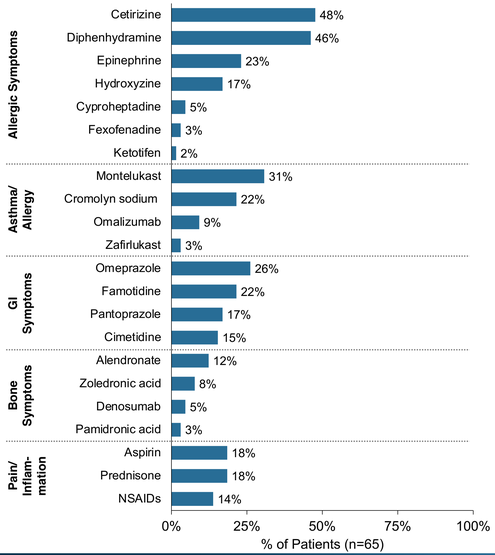
Figure 3. Signs and Symptoms at First Visit to Physician/Upon Diagnosis



- The distribution of symptom-directed therapies prescribed to ISM patients are reported in Figure 4.
- 95% of patients were receiving more than one treatment to manage their ISM (range 1-9).

Figure 4. Treatments Prescribed to Patients for ISM

Note: Patients could have been prescribed multiple treatments.



Conclusions

- Patients with ISM present with multiple signs and symptoms and co-morbid conditions and are referred by a wide range of specialties.
- Increasing the awareness of heterogeneous signs and symptoms of ISM may help improve the recognition of the disease and decrease the time to diagnosis.
- Patients receive a wide array of symptom-directed therapies to manage their ISM.

References

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3. Jennings SV, Slee VM, Zack RM, et al. Patient Perceptions in Mast Cell Disorders. *Immunol Allergy Clin North Am.* 2018;38(3):505-525. doi:10.1016/j.iac.2018.04.006

Disclosures

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