Suggestions for the monitoring and follow up of patients with systemic mastocytosis based on a single centre tertiary experience.

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Introduction

There are no agreed guidelines for the monitoring and follow up of patients with systemic mastocytosis (SM). We feel that suggestions on guidance in this area would be beneficial to patient care.

Methods

We undertook a review of our cohort of SM patients to determine trends in their follow up to help develop suggestions in this area. Patients were identified from clinics between May 2018 - April 2019. Hospital and laboratory records were used to collect demographic details, blood results (including molecular profiles), bone mineral densitometry (BMD) and abdominal ultrasound (US) results. Serial mast cell tryptase (MCT) levels were collected with a minimum interval of 3 months between results. The trends and outcomes were documented and analysed to formulate suggestions on the monitoring and follow up these patients.

Cohort

- 22 patients (male= 9, female =13)
- Median age of 59 years (range 28- 90 years)
- Their diagnoses (as per WHO classification[1]) were:
    - Indolent SM (n= 13)
    - SM with associated haematological neoplasm (AHN) (n=3)
    - Smouldering SM (n=2)
    - Aggressive SM (n= 2)
    - Pre-diagnostic SM (n=2)- where the WHO criteria for SM were not fully met [2].
- Median MCT levels at diagnosis: 50.0ng/ml (range 6.3ng/ml - 550ng/ml).
- c-KIT mutations
    - 17/22 positive (D816V n= 16, D816Y n=1).
    - 2/22 negative
    - Results unavailable for 3/22
- Cutaneous involvement was seen in 16/22 patients.

Follow-up data

- Follow-up data was available for 20/22 patients.
- Median follow up of 58.9 months (range 7.7 to 178.3 months).
- Average change in MCT levels was an increase of 1.7%
    - Average interval of 255 days between measurements
- US assessment:
    - 17/20 patients had a baseline US
    - 1 patient found to have radiological splenomegaly
    - Of the 16 patients who had a normal US result at baseline, 9 patients went on to have serial scans (total number of scans per patient range 2-3).
    - Of these 0 patients went on to develop radiological splenomegaly
- BMD:
    - 18/20 patients had baseline BMD recorded.
    - 6/18 patients were found to be osteopenic and 3/18 were osteoporotic.
    - Of the 9 patients with normal BMD scans initially, 5 had serial scans (total number of scans per patient range 2-4). Of these, no patients developed osteopenia or osteoporosis.
- Pre procedural prophylaxis:
    - In our cohort 3/20 patients had procedures requiring general anaesthetic.
    - They were given a standard preoperative prophylaxis regime (see figure 2) to minimise the risk of anaphylaxis without any adverse outcomes.
- Epipens:
    - 4/20 patients had a history of anaphylaxis. 5/20 patients had repeat prescriptions for Epi-pens. Of the 15/20 patients without regular Epipens, there were no episodes of anaphylaxis during their follow up.

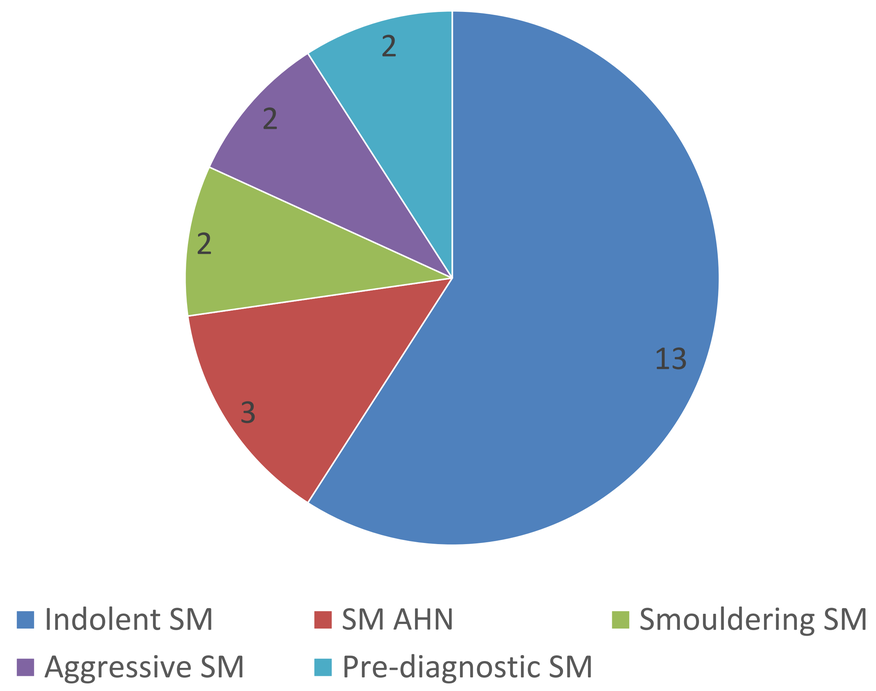

Figure 1: Diagnosis

Timing	Medication
24 hours pre-procedure	Prednisolone 50mg stat with PPI
1 hour pre-procedure	Prednisolone 50mg stat with PPI Fexofenadine 120mg stat Ranitidine 150mg stat Monteleukast 10mg stat

Figure 2: Pre-procedure prophylaxis

Suggestions for management

Based on local experience, we offer the following suggestions for the management of patients with stable SM:

1. All patients should have a baseline clinical assessment including examination for organomegaly (especially liver and spleen size).
2. Baseline standard bloods including MCT levels.
3. Baseline abdominal US for spleen size and BMD should be performed.
4. Standard annual review (shortened to 6 monthly at clinician's discretion) with clinical assessment and standard bloods including mast cell MCT levels.
5. Interval US should only be performed if clinically indicated.
6. BMD to be performed 3 yearly.
7. Epi-pen if *any* significant mast cell mediator symptoms or history of allergy/ anaphylaxis.
8. Standard protocol for pre-procedural anaphylaxis prophylaxis protocol should be followed (see figure 2).

References

[1] Valent, P., Akin, C. and Metcalfe, D., 2017. Mastocytosis: 2016 updated WHO classification and novel emerging treatment concepts. *Blood*, 129(11), pp.1420-1427.
[2] Pardanani, A., 2018. Systemic mastocytosis in adults: 2019 update on diagnosis, risk stratification and management. *American Journal of Hematology*,.