



Idiopathic anaphylaxis and systemic mastocytosis

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A 51-year-old Caucasian male presents with history of idiopathic anaphylaxis and gastrointestinal symptoms including diarrhea, abdominal pain and dyspepsia. He reported delayed and immediate reactions to red meat, and allergic rhinitis symptoms when exposed to cats. CBC and BMP were within normal range, except for IgE 150 IU/ml (reference: 0–90 IU/ml) and tryptase 25 ng/ml (reference: < 11.5 ng/ml). Bone marrow biopsy was performed to evaluate for mastocytosis. Figure 1 shows H&E of a normocellular marrow with a nodular infiltrate (panel a, 40 ×) consisting of aggregates of atypical mast cells (> 15 cells) (panel b, 400 ×), admixed with lymphocytes and eosinophils (panel c, 1000 ×). Immunohistochemistry revealed mast cells positive for CD117 (panel d, 400 ×), tryptase (panel e, 400 ×), CD25 (panel f, 400 ×), variable CD2 (panel g, 400 ×), and dim CD30 (panel h, 400 ×). Aspirate smear showed atypical morphology of mast cells including spindled (panel i, 1000 ×) and

degranulated forms (not shown). Flow cytometry demonstrated cells positive for CD117 (bright) (not shown) with aberrant expression of CD25 and CD2 (spectrum) (panel j). In addition, real-time PCR detected KIT^{D816V} mutation. No evidence for an associated hematological neoplasm was present. Altogether, these findings fulfill the criteria for systemic mastocytosis (SM) according to the WHO 2016 guidelines. This presentation underscores the importance of complete investigation for clonal mast cell disorders in patients with idiopathic anaphylaxis and highlights the indolent course of SM in a subpopulation of patients [1].

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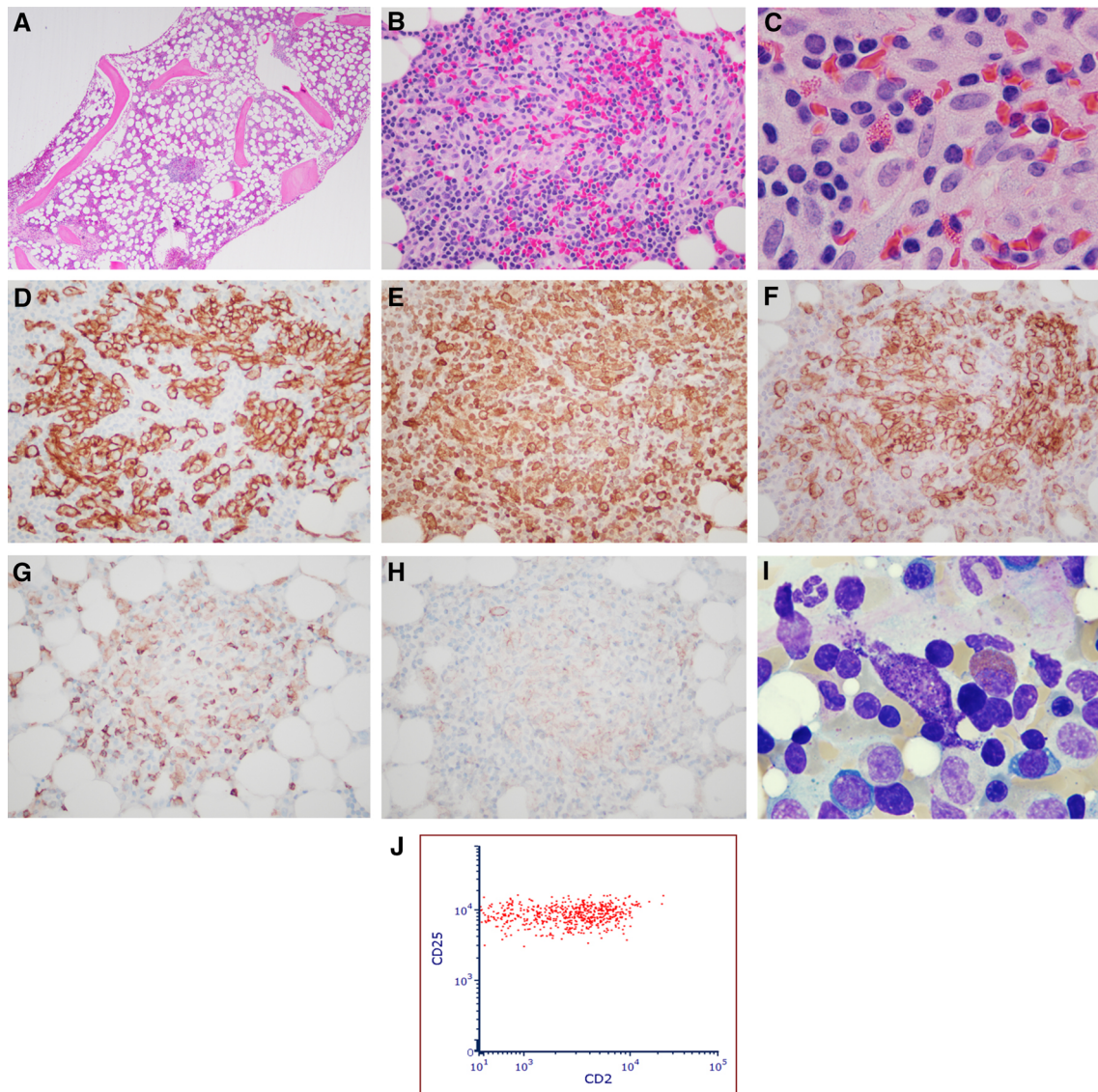


Fig. 1 Nodular aggregates of mast cells involving the marrow on H&E (a–c). Immunohistochemistry shows mast cells positive for CD117 (d), tryptase (e), CD25 (f), variable CD2 (g), and dim CD30

(h). Aspirate smear with a spindle-shaped neoplastic mast cell is depicted in (i). Flow cytometry reveals mast cells with aberrant expression of CD2 and CD25 (j)

Compliance with ethical standards

Conflict of interest The authors have no conflict of interest.

References

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