Anaphylaxis and systemic mastocytosis caused by *Solenopsis invicta* stings

*Anafilaxia e mastocitose sistêmica ocasionada pela* *Solenopsis invicta*

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Indolent systemic mastocytosis is a rare disease characterized by an increased number of mast cells in the bone marrow and other tissues, such as the liver, spleen, lymph nodes, and skin. Patients with indolent systemic mastocytosis and high serum tryptase levels are at risk for Hymenoptera venom-induced anaphylaxis. Hymenoptera venom immunotherapy in patients with specific IgE is safe and effective. While some patients can receive ultra-rush venom immunotherapy with minimal side effects, omalizumab effectively protects against anaphylaxis during the build-up phase.

**Keywords:** Anaphylaxis, indolent systemic mastocytosis, urticaria pigmentosa, imported fire ant, *Solenopsis invicta*, hymenoptera venom immunotherapy, hereditary alpha-tryptasemia.

Indolent systemic mastocytosis (ISM) is a rare disease characterized by an increased number of mast cells (MCs) in the bone marrow (BM) and other tissues, such as the liver, spleen, lymph nodes, and skin, and a normal life span. Skin lesions associated with ISM are typically maculo papular monomorphic lesions, also known as urticaria pigmentosa (UP). When UP lesions are stroked, a wheal and flare reaction is noted within a few minutes, known as Darier’s sign.¹-³ Patients with ISM and high serum tryptase levels are at risk for Hymenoptera venom-induced anaphylaxis, which is more common in males. These patients with Hymenoptera venom-specific IgE are candidates for immunotherapy, which is recommended for life, and is effective at protecting most patients from future severe anaphylactic episodes.⁴ ⁵ We present here the first case of life-threatening anaphylaxis following multiple stings from imported fire ants (IFA) in a female as the presenting symptoms leading to the diagnosis of ISM.⁶
Case report

A 31-year-old caucasian woman presented with a long history of perennial allergic rhinitis and atopic dermatitis as a child. While outdoors and barefoot, she was stung for the first time by multiple IFA in her legs and feet. She immediately developed throat tightening with difficulty breathing, and abdominal cramping pains, became hypotensive, and developed grand mal seizures with fecal and urine incontinence. She was resuscitated with three IM 0.3mg epinephrin injections, promethazine 50mg IV (H1-antihistamine), hydrocortisone 300mg IV, replacement fluids, and oxygen, recovering within a few hours without sequelae. She tested positive in prick skin testing and serum-specific IgE to IFA *Solenopsis invicta* (Si) (0.47 kU/L; negative value below 0.10 kU/L), house dust mites (*Dermatophagoides farinae*, *Dermatophagoides pteronyssinus*, and *Blomia tropicalis*), dogs, cats, and horses. Blood cell counts, platelets, liver, and renal function tests were all normal. Specific IgE to honey bee, wasp and hornets were all negative.

Serum tryptase levels were 22.8 and 23.7 ng/mL at baseline (normal value below 11.4 ng/mL). Peripheral blood KIT mutations in exons 8 and 17 were not detected. A bone marrow biopsy was obtained and showed a negative KIT D816V mutation. The histology presented aggregates of 15 or more MCs stained by tryptase, CD117 spindle-shaped forms, and aberrant expression of CD25, negative for CD2, CD3, CD30, and CD34. An abdominal CT scan was negative for hepatosplenomegaly, and a DEXA scan showed osteopenia. The diagnosis of ISM was established. The patient developed photo-allergic dermatitis to a solar protection cream containing parabens with a generalized flare of UP lesions with a positive Darier’s sign (Figure 1). Genetic testing showed a normal alpha-tryptase copy number 1 for the gene TPSAB1 (GENEbyGENE, Houston, TX), ruling out Hereditary alpha tryptasemia (HαT). Her medications included: 20mg H1-antihistamine bilastine, 400mg H2-antihistamine cimetidine, 10mg montelukast, calcium, and vitamin D. Fluticasone furoate nasal spray was prescribed for allergic rhinitis. During acute episodes of UP flares, the patient uses 40mg prednisolone and topical 0.1% tacrolimus ointment. She carries three 0.3mg epinephrine autoinjectors all the time. The patient was treated with immunotherapy with IFA-whole body extract, and has achieved monthly maintenance dosing with 0.5mL of 1:100 wt/vol Si (Greer, Lenoir, North Carolina). No more episodes of anaphylaxis have occurred for over a year.

Discussion

This case illustrates the need to obtain baseline serum tryptase measurement in all patients with Hymenoptera venom anaphylaxis to screen for mast cell activation disorders. A value above the normal range should prompt the determination of KIT D816V mutation in peripheral blood and a bone marrow biopsy. Severe anaphylaxis has also been associated with HαT, with duplication of TPSAB1 alpha-tryptase gene. In this case the TPSAB1 copy number analysis was normal, ruling out HαT. Hymenoptera venom immunotherapy in patients with specific IgE is safe and effective. While some patients can receive ultrarush venom immunotherapy with minimal side effects, omalizumab has effectively protected against anaphylaxis during the build-up phase.

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References


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