

Mastocytosis with Bee Sting Anaphylaxis

Dennis K. Ledford, MD



This feature presents highlights from the Ask the Expert section of the American Academy of Allergy, Asthma & Immunology (AAAAI) Web site written by the Ask the Expert editors. For more questions and answers, visit www.aaaai.org/ask-the-expert.aspx.

Question:

A 76-year-old white man with confirmed mastocytosis (serum tryptase levels > 150 ng/mL for last 7 years and D816V (c-KIT mutation) presents with syncope 15 minutes following a bee sting in his foot. He was taken to a hospital emergency department, resuscitated, and discharged without admission. Past medical history is notable for chronic sinus issues and penicillin allergy but no asthma. Mastocytosis is indolent and clinically stable with only pruritis (patient is receiving 10 mg loratidine twice a day) and he has an epinephrine autoinjector. What is the best option and recommendation for this patient?

Answer:

Thank you for your question.

Patients with mastocytosis, as well as mast cell activation disorders, are at increased risk of anaphylaxis following insect stings. Although they are also at greater risk of reaction to allergen immunotherapy, this would be the desirable therapy because the risk of a reaction to supervised immunotherapy is preferable to unsupervised field stings. You should document a discussion emphasizing the increased risk for anaphylaxis in general and the need to keep an epinephrine autoinjector readily available. We all feel apprehension about the use of epinephrine in older individuals, but any increase in risk is less than the risk of untreated anaphylaxis and hypotension. You do not mention, but I would

recommend that beta-blocker therapy be avoided, if possible, and I would also prefer that angiotensin-converting enzyme inhibitors be avoided. Angiotensin receptor blockers may also interfere with the physiologic response to hypotension so I would ask that these agents not be used unless specific need is documented.

I have copied some excerpts from the Practice Parameter on Insect Allergy below.¹ I also provided a reference from the *Journal of Allergy and Clinical Immunology: In Practice* that you may find of interest.²

In contrast, patients with a history of anaphylaxis to a sting have an average of almost 50% frequency of systemic reaction to a sting. Patients with large local reactions have less than 10% chance of a systemic reaction (and less than 5% chance of anaphylaxis). However, no test predicts the severity of a sting reaction (other than basal serum tryptase).

Compared to other causes of anaphylaxis such as foods or medications, the prevalence of mast cell disorders is higher in patients who have had anaphylaxis to an insect sting. Therefore, measurement of basal serum tryptase should be considered in all patients who are candidates for venom immunotherapy (VIT). Elevated basal serum tryptase is closely correlated with the risk of severe anaphylaxis to stings, and is most frequently found in patients with reactions including hypotensive shock. The frequency of abnormal basal tryptase is much lower in patients with less severe systemic reactions to stings, and the clinical significance in these patients is less clear. There is a cost and burden associated with abnormal results of basal tryptase (eg: bone marrow biopsy, consultation with other specialists, anxiety associated with an abnormal test result). However an abnormal result is associated with severe anaphylaxis to stings, increased risk of systemic reactions during VIT (to a sting or venom injection), and greater risk of sting anaphylaxis after stopping VIT. With these considerations in mind, measurement of basal serum tryptase is highly recommended in patients who had hypotensive reactions to a sting, and should be considered in other patients with systemic reactions to stings. Additionally, elevated basal tryptase may indicate the presence of an occult mast cell disorder and may be present in sting-allergic patients with negative venom allergy tests as well.

Encourage continuation of VIT for an extended time, or indefinitely, in patients with high-risk factors, such as very severe reaction before VIT (syncope, hypotension, severe respiratory distress), systemic reaction during VIT, honeybee allergy, and increased basal serum tryptase levels. (Strong Recommendation; C evidence).¹

University of South Florida, Morsani College of Medicine, Tampa, Fla

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College of Medicine and the Veterans Administration; has provided expert witness testimony for Wicker, Smith, O'Hara, McCoy, and Ford on the topic of asthma death; has provided expert witness testimony for Shook Hardy Bacon on the topic of drug allergy; has provided expert witness testimony for Saieva and Stine on the topic of metal allergy; has provided expert witness testimony for Vedrine Associates on the topic of drug allergy; has received consultancy fees from Genentech and AstraZeneca; has received Speaker's fees from TEVA, Boehringer Ingelheim, Novartis, AstraZeneca, Genentech, and Sanofi/Regneron; has received research support from AstraZeneca; is on the Editorial Board for *Annals of Allergy Asthma and Immunology*; is coeditor of *Asthma: Co-Morbidities, Co-existing Conditions and Differential Diagnosis*; is editor, *Allergens and Allergen Immunotherapy*; is volume editor, *Encyclopedia of Allergy, Immunology and Vaccines*; is coeditor, *Asthma and Allergy Basics*; and is an UpToDate contributor.

Corresponding author: Dennis K. Ledford, MD, 13000 Bruce B Downs Blvd, VAR111D, Tampa, FL 33613. E-mail: dledford@health.usf.edu.

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In summary, the tryptase elevation of mastocytosis or other mast cell disorders is a marker for risk of more severe insect allergy and identifies need for treatment and longer-term venom or imported fire ant whole-body extract immunotherapy.

I hope this information is of help to you and your practice.

REFERENCES

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