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Autor(en) des Beitrags: Brockow, K; Jofer, C; Behrendt, H; Ring, J

Titel des Beitrags: Anaphylaxis in patients with mastocytosis: a study on history, clinical features and risk factors in 120 patients.

Abstract: BACKGROUND: Excessive mast cell mediator release may lead to anaphylaxis in patients with mastocytosis. However, the incidence, clinical features and trigger factors have not yet been analyzed. METHODS: To identify risk factors for anaphylaxis in mastocytosis, we determined cumulative incidence, severity, clinical characteristics, and trigger factors for anaphylaxis in 120 consecutive patients (53 male; 67 female, median age and range 24 years, 1 month to 73 years), and correlated these with disease severity of mastocytosis, skin involvement, basal total serum tryptase, and diaminooxidase concentrations. RESULTS: The cumulative incidence of anaphylaxis in patients with mastocytosis was higher in adults (49%; P< 0.01) compared with that in children (9%). Only children with extensive skin involvement had experienced anaphylaxis. In adults, anaphylaxis was correlated to the absence of urticaria pigmentosa lesions (P< 0.03). Reactions occurred more frequently in adults with systemic (56%) when compared with cutaneous mastocytosis (13%; P< 0.02). In adults, 48% of reactions were severe, and 38% resulted in unconsciousness. Major perceived trigger factors for adults were hymenoptera stings (19%), foods (16%), and medication (9%); however, in 26% of reactions, only a combination of different triggers preceded anaphylaxis. Trigger factors
remained unidentified in 67% of reactions in children compared with 13% in adults. Patients with anaphylaxis had higher basal tryptase values (60.2 +/- 55 ng/ml, P< 0.0001) in comparison with those without (21.2 +/- 33 ng/ml), but not diaminooxidase levels. CONCLUSION: Adult patients and children with extensive skin disease with mastocytosis have an increased risk to develop severe anaphylaxis; thus, an emergency set of medication including epinephrine is recommended.