A study on serum IgE and clinical symptomatology of atopy in patients suffering from the lysosomal storage disorder Fabry disease.

BACKGROUND: Lysosomal storage disorders may impair intracellular lysosomal processing of antigen with consequences for antibody production [e.g. immunoglobulin E (IgE)] and atopic disease status. AIMS: Serum concentration of total IgE as well as clinical symptoms of atopic disorders as an indirect consequence of lysosomal impairment of antigen processing were studied in male and female Fabry disease (FD) patients with and without replacement of the missing lysosomal enzyme, alpha-galactosidase A (alpha-Gal A).

METHODS: Observational study in 31 adult FD patients with measurements of total serum IgE concentration. Questionnaire-derived data were obtained for atopic eczema (AE) skin lesions, allergic rhinoconjunctivitis (RCA) and allergic asthma (AA) at present or in the past.

RESULTS: Among 12 FD males under enzyme replacement therapy (ERT), 2 showed total IgE concentrations above 100 kU/L. Clinical symptoms for AA were found in 2, RCA and AE in 1, respectively. Among 10 FD females under ERT, 4 showed total IgE concentrations above 100 kU/L. Clinical symptoms for AA were found in 4, RCA in 2 and AE in 2. Among 9 females without ERT, 2 showed total IgE concentrations above 100 kU/L. Clinical symptoms for AA were found in 2, RCA in 2 and AE in none.

CONCLUSIONS: FD patients may demonstrate an increased total serum IgE concentration and may show symptoms of atopic disorders (AA, ...
RCA, AE) in a prevalence rate comparable to international experience for individuals without FD. There was no difference between patients with and without ERT. Lack of detection of AE in females without ERT is suggested to be caused by the small sample size. FD patients without any alpha-Gal A activity prior to initiation of ERT should be in the focus of future studies.

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