Abnormal T-cell phenotype in episodic angioedema with hypereosinophilia (Gleich's syndrome): frequency, clinical implication and prognosis

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Résumé en anglais	BACKGROUND: Episodic Angioedema with eosinophilia (EAE, Gleich's syndrome) is a rare disorder consisting of recurrent episodes of angioedema, hypereosinophilia and frequent elevated serum Immunoglobin M. METHODS: We conducted a retrospective multicenter nationwide study regarding the clinical spectrum and therapeutic management of patients with EAE in France. RESULTS: Thirty patients were included with a median age at diagnosis of 41 years [5-84]. The median duration of each crisis was 5.5 days [1-90] with swelling affecting mainly the face and the upper limbs. Total serum IgM levels were increased in 20 patients (67%). Abnormal T-cell immunophenotypes were detected in 12 patients (40%) among which 5 (17%) showed evidence of clonal TCR γ gene rearrangement. Median follow-up duration was 53 months [31-99]. The presence of an abnormal T-cell population was the sole factor associated with a shorter time to flare (hazard ratio 4.15 [CI 95% 1.18-14.66; p=0.02). At last follow-up, 3 patients (10%) were able to withdraw all treatments and 11 (37%) were in clinical and biological remission with less than 10 mg of daily prednisone. CONCLUSION: EAE is a heterogeneous condition that encompasses several disease forms. Although patients usually respond well to glucocorticoids, those with evidence of abnormal T-cell phenotype have a shorter time to flare.
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