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Introduction: Hereditary angioedema resulting from the deficiency of the C1-inhibitor (HAE-C1-INH) is an autosomal dominant disorder. It is characterized by episodic recurrences of bradykinin-mediated edema formation in the subcutis and/or the submucosa. C1q determination has differential diagnostic value in distinguishing between the hereditary and the acquired forms of C1-INH deficiency. Although the C1q levels of the HAE-C1-INH patients are usually within the normal range, and the presence of anti-C1q autoantibodies is not characteristic either, both were abnormal in 13% of our cases. Our objective was to investigate C1q and anti-C1q levels in HAE-C1-INH.

Methods: The study population comprised 141 patients with HAE-C1-INH (80 females and 61 males, mean age: 37 years (min: 3, max: 82), 130 with HAE type I, and 11 with HAE type II. We measured C1q, anti-C1q, C4, functional and antigenic C1-INH levels and then, recorded the number and location of edematous episodes, as well as monitored drug therapy over the subsequent year.

Results: C1q correlated with C4 and functional C1-INH levels, as well as – in patients with HAE type 1 – with antigenic C1-INH level ($r=0.2154$, $p=0.0106$; $r=0.1966$, $p=0.0195$; and $r=0.2413$, $p=0.0079$). Anti-C1q unrelated to all the other complement parameters in all patients. Although C1q was not related to the number of edematous episodes either, anti-C1q level showed a positive relationship with the number of laryngeal edema attacks ($r=0.2463$, $p=0.0040$). Analyzing female and male patients separately, we found that anti-C1q correlated with total attack number ($r=0.2783$, $p=0.0143$), as well as with the number of submucosal (laryngeal and abdominal) episodes ($r=0.2834$, $p=0.0125$, and $r=0.2317$ $p=0.0426$) in women only. The comparison of patients treated/not treated with danazol revealed a relationship between anti-C1q level and facial edema in danazol-treated patients, as well as a correlation between the former and laryngeal edema patients not receiving danazol ($r=-0.3708$, $p=0.0132$; $r=0.2232$, $p=0.0377$).

Discussion: As indicated, in particular, by the clinical relationships found in females and in patients with more severe disease (requiring treatment with danazol), the presence of anti-C1q autoantibodies might influence the clinical situation or might be a clinical marker even if their level is normal. The lack of a similar correlation with C1q suggests immunoregulatory relationships in the pathomechanism of HAE-C-INH, rather than activation triggering the classical pathway.