ELEVATED NEUTROPHIL ELASTASE LEVELS IN HEREDITARY ANGIOEDEMA

Nóra Veszeli
Nóra Veszeli, Dorottya Csuka, Zsuzsanna Zotter, Éva Imreh, Szabolcs Benedek, Varga Lilian, Farkas Henriette

3rd Department of Internal Medicine, Semmelweis University, Budapest

Introduction: Hereditary angioedema due to C1-inhibitor deficiency (C1-INH-HAE) is a rare autosomal dominant disorder. It is characterised by the episodic swelling of subcutaneous tissues, the intestinal wall, and the upper airway mucosa.
According to several case reports and to our recent studies, white blood cell count may increase (mostly of neutrophil granulocytes [NG]) during attacks of C1-INH-HAE. The production of neutrophil elastase (NE) – as a marker of NG activation- during oedematous attacks has not yet been investigated. As shown by earlier studies, NE can inactivate C1-INH. This might contribute to the activation of the plasma enzyme systems inhibited by C1-INH, and thereby also to oedema formation. Our questions were whether NE level is elevated in C1-INH-HAE patients in symptom-free periods and during attacks, as well as whether NE is correlated with NG count or functional C1-INH activity.
Methods: We determined the complete blood count and leukocyte differential in blood samples collected during and between oedematous episodes from 26 C1-INH-HAE patients, as well as from 26 healthy controls. We also measured the serum levels of NE and of functional C1-INH.
Results: Both WBC and NG were significantly elevated (after adjustment for haemoconcentration) in samples obtained during attacks compared with those collected in a symptom-free period ($p=0.0184$ and $p=0.0419$ respectively). In agreement with our earlier findings, these parameters were lower in healthy controls than in patients either during or between attacks ($p<0.0001$, $p=0.0261$ and $p=0.0004$, $p=0.0458$). In C1-INH-HAE patients, NE levels were higher in samples obtained during an attack than in those collected from the same patients during a symptom-free period ($p=0.0350$). In healthy controls, NE levels were lower than the concentration measured in patients either during or between attacks ($p=0.0004$, $p=0.0101$,). We did not find a correlation between NE level and NG count or between the former and functional C1-INH activity.
Conclusion: Patients with C1-INH-HAE have elevated NG counts and NE levels; both increase further during attacks. Nevertheless, NE level does not correlate either with NG count or with functional C1-INH activity. This might suggest that systemic neutrophil activation and the attenuation of C1-INH function are both uncharacteristic. This, however, does not exclude the local activity of other pathophysiological processes at the site of oedema formation.

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