Angioedema with normal C1 Inhibitor

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The emergence of clinical angioedema (AE) is subsequent to bradykinin (BK) production, with AE attacks resulting from a local endothelial permeability in the affected tissue. Kallikrein-kinin system (KKS) and BK generation mediate this process.
AE with normal C1Inhibitor (C1Inh) is difficult to diagnose. It is known to be unresponsive to treatment with antihistamines, corticosteroids, and epinephrine. AE with normal C1Inh is divided between patients with a known *F12* mutation, and recently with Plasminogen or angiopoietin mutations, and those with unknown origin [1].

However, when the patient presents with an acute crisis, the biological profile is of great support for the decision of treatment and follow-up.

Here, we want to describe 4 patients admitted at the acute phase of angioedema in emergency department: abdominal cramps, respiratory distress, AE of the eyelids, tongue and lips. The diagnosis was often delayed. Despite the fact that one of the patients needed tracheal intubation in the past, he suffered from recurrent AE, without diagnosis, for the last 25 years. We believe that KKS biological workup is helpful. Three samples were harvested at different times during the attack and investigated. C1 Inhibitor (C1Inh) function, spontaneous amidase activity, kinin measurement and kininogen cleavage were analyzed as described previously [2,3,4,5,6].

The results demonstrate that KKS activation, and subsequently BK production, are the key actors of these attacks, without decrease of C1Inh function. They also demonstrated a very important increased kinin forming process with increased spontaneous amidase activity in line with kininogen (HK) cleavage.
In addition, the administration C1Inh concentrate results in clinical improvement.

The symptomatology of angioedema is subsequent to the enzymatic activity responsible for kinin production. Kinetics of biological events demonstrates KKS activation and BK production. Laboratory confirmation using KKS biological parameters seems to be helpful for diagnosis and patients management.

References:

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