Bradykinin-mediated angioedema: biological diagnosis

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Bradykini-mediated angioedema (BK-AE) is classically non-pruritic, non-erythematous swelling affecting submucosal or subcutaneous tissues. AE could be life-threatening as soon as affecting the larynx. AE attack is the consequence of kallikrein-kinin system (KKS) activation with subsequent bradykinin (BK) accumulation onto the endothelium. BK is released from the cleavage of high molecular weight kininogen (HK) cleavage after kallikrein (PKa) activation. The regulation of KKS is mainly assured by C1 inhibitor (C1Inh).
Bradykinin-mediated angioedema is difficult to diagnose, 50% of patients with Hereditary angioedema have previously had their conditions misdiagnosed. The challenge of the Bradykinin-mediated angioedema diagnosis by the rarity and the heterogeneity of its affected population, and the overlap in symptoms with other more common diseases.
Misdiagnosis results in marked delays in receiving the correct diagnosis, during which time. The median of this delay is 8.5 years, the mean delay to diagnosis can be up to 16 years, even in developed countries.

Bradykinin mediated angioedema results in from a loss of control of KKS, from overproduction or failure to inactivate the BK. The biological approach is based on angioedema pathophysiology, it explores KKS biological parameters: KKS activation to demonstrate the kinin forming, the control of KKS which is investigated by the functional activity of C1Inh and kinin catabolism to measure the activities of BK catabolism enzymes.

The biological profile is of great support for the decision of treatment and follow-up.