Figure1. Proposed pathophysiological mechanisms in hereditary α-tryptasemia (HαT). In comparison with normal (wild-type, WT) mast cells (MC), those from individuals with HαT possess extra *TPSAB1* copies resulting in a more significant formation of α/β heterotetramers. Following MC degranulation, α/β heterotetramers are released, and might activate epidermal growth factor (EGF)-like module-containing mucin-like hormone receptor-like 2 (EMR2) and protease-activated receptor 2 (PAR2), inducing a decreased threshold for vibration-induced mast cell degranulation, and increasing vasopermeability and bronchial/gastrointestinal smooth muscle contraction, respectively, explaining manifestations commonly found in patients with HαT (e.g., urticaria, abdominal pain and diarrhea).