



Fig. S1. Two female patients, aged (A) 60 and (B) 10 years with undisputable C1INH-HAE type 1; low C1Inh function and *SERPING1* gene mutations (A) c.889+1delG and (B) c.1480C>T. Both presented with acute pitting urticaria as a prodromal manifestation of attacks of hereditary angioedema (HAE). Oedematous and papular characteristics of the rash were obvious and suggestive of urticaria. Skin biopsy was not available.