



Fig. S1. (a) Clinical features on the abdomen of patient 1. (b) Indirect immunofluorescence (IF) using 1M NaCl-split skin in patient 1 ($\times 100$). (c) Clinical features on the left dorsal foot in patient 2. (d) Indirect IF using 1M NaCl-split skin in patient 2 ($\times 100$). (e) Results of immunoglobulin G (IgG) immunoblotting using normal human epidermal extract. Control pemphigus vulgaris serum (PV) showed the 160 kDa desmoglein 1 and the 130 kDa desmoglein 3 (*lane 1*), control paraneoplastic pemphigus serum (PNP) showed the 210 kDa envoplakin, the 190 kDa periplakin, and control bullous pemphigoid (BP) serum showed 230 kDa BP230 and 180 kDa BP180 (*arrows on the left*). *Lanes 4 and 5*: sera of patients 1 and 2, respectively. (f) Immunoblotting using recombinant proteins (RP) of monomeric NC16a domain of BP180. Control BP serum (*lane 1*), but not normal serum (*lane 2*), reacted with this RP (*arrow*). *Lanes 3 and 4*: sera of patients 1 and 2, respectively. (g) Immunoblotting using RP of BP180 C-terminal domain. Control BP180-type mucous membrane pemphigoid (MMP) serum (*lane 1*), but not normal serum (*lane 2*), reacted with this RP (*arrow*). *Lanes 3 and 4*: sera of patient 1 and patient 2, respectively. *Non-specific reactivity.