Appendix S1

Histology and immunodetection. After informed consent was obtained, punch biopsies of lesional skin (cutis laxa without urticaria) of the hands were taken, snap-frozen and stored at –80°C. All the stainings were performed by routine techniques at the department of anatomo-pathology. IgA deposits were revealed with FITC-conjugated anti-human IgA antibodies (Jackson ImmunoResearch, Suffolk, UK).

Electron microscopy and immunoelectron microscopy. The biopsy specimen was immersed in Sorensen's buffer (0.1M, pH 7.4) with 2.5% glutaraldehyde overnight at 4°C, post-fixed in 0.5% osmic acid, dehydrated and embedded in EmBed 812 using an Automated Microwave Tissue Processor for Electronic Microscopy, Leica EM AMW. Thin sections (70 nm; Leica-Reichert Ultracut E) were collected at several levels of each block and counterstained with uranyl acetate. For immunodetection, biopsies were embedded at –30°C in lowicryl HM-20 (EMS, Hatfield, PA) in a Leica AFS (automatic freeze-substitution instrument) and polymerised upon ultraviolet light. Lowicryl ultrathin sections were collected on Foamvar carbon-coated nickel grids and processed for immunodetection as follows: incubation with anti-human IgA antibody overnight at 4°C, incubation with colloidal gold-conjugated anti-goat antibody (Aurion, France), and post-fixation with 1% glutaraldehyde in PBS. Negative controls were performed by omitting the primary antibody.

Western blot. Lesional dermal fibroblasts (HDF) were obtained from punch biopsy using the explant technique Protein extracts and protein analysis were done as described previously (10) with anti-fibulin-5 antibodies purchased from Santa Cruz, Heidelberg, Germany.

Mutation analysis of fibulin-5 (FBLN5) and elastin (ELN) genes. DNA extraction from EDTA blood samples was performed using standard procedures. After PCR amplification, molecular analysis of the *FBLN5* and *ELN* genes was performed via direct sequencing of the coding region and the intron/exon boundaries using a 3730xl Sequencer (Applied Biosystems®). The data were analysed using Seqscape software (Applied Biosystems®).