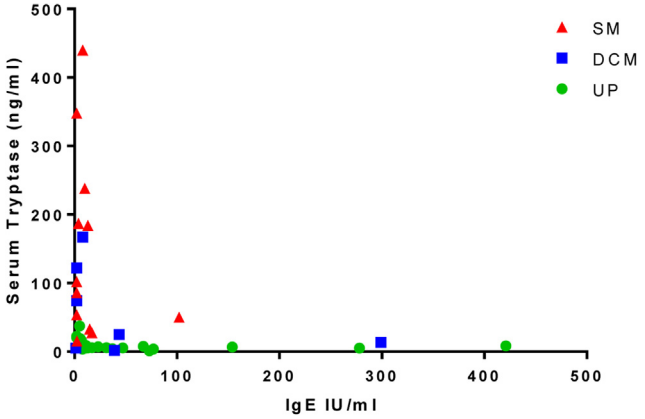

FIG E1. Serum tryptase levels (in nanograms per milliliter) versus serum IgE levels (in international units per milliliter) are shown. Patients with UP are represented by *green dots*, patients with DCM are represented by *blue squares*, and patients with ISM are represented by *red triangles* (Spearman rho = 0.517, $P < .0001$).

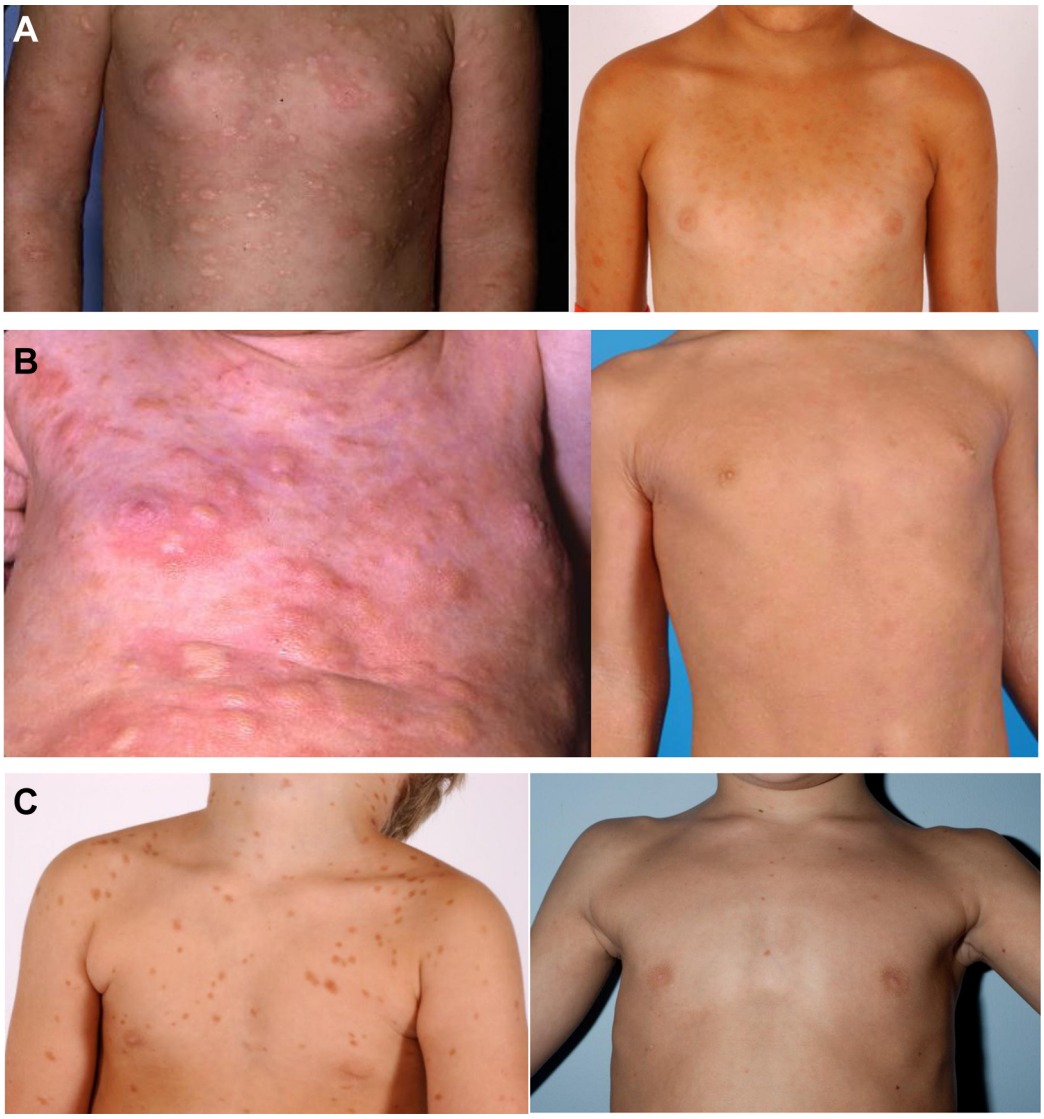

FIG E2. Appearance of skin lesions over time with concurrent serum tryptase levels. Regression of skin lesions in patients with UP, predominately maculopapular cutaneous mastocytosis lesions with some nodular lesions that evolve into a strictly maculopapular cutaneous mastocytosis (ages 2 and 7 years; tryptase level, 20 and 6.6 ng/mL, respectively; **A**); DCM (ages 17 months and 9 years; tryptase levels, 17.4 and 4.9 ng/mL, respectively; **B**); and ISM with a typical maculopapular cutaneous mastocytosis pattern (ages 3 and 9 years; tryptase levels, 32.9 and 12.6 ng/mL, respectively; **C**) are shown and are consistent with decreases in serum tryptase levels.

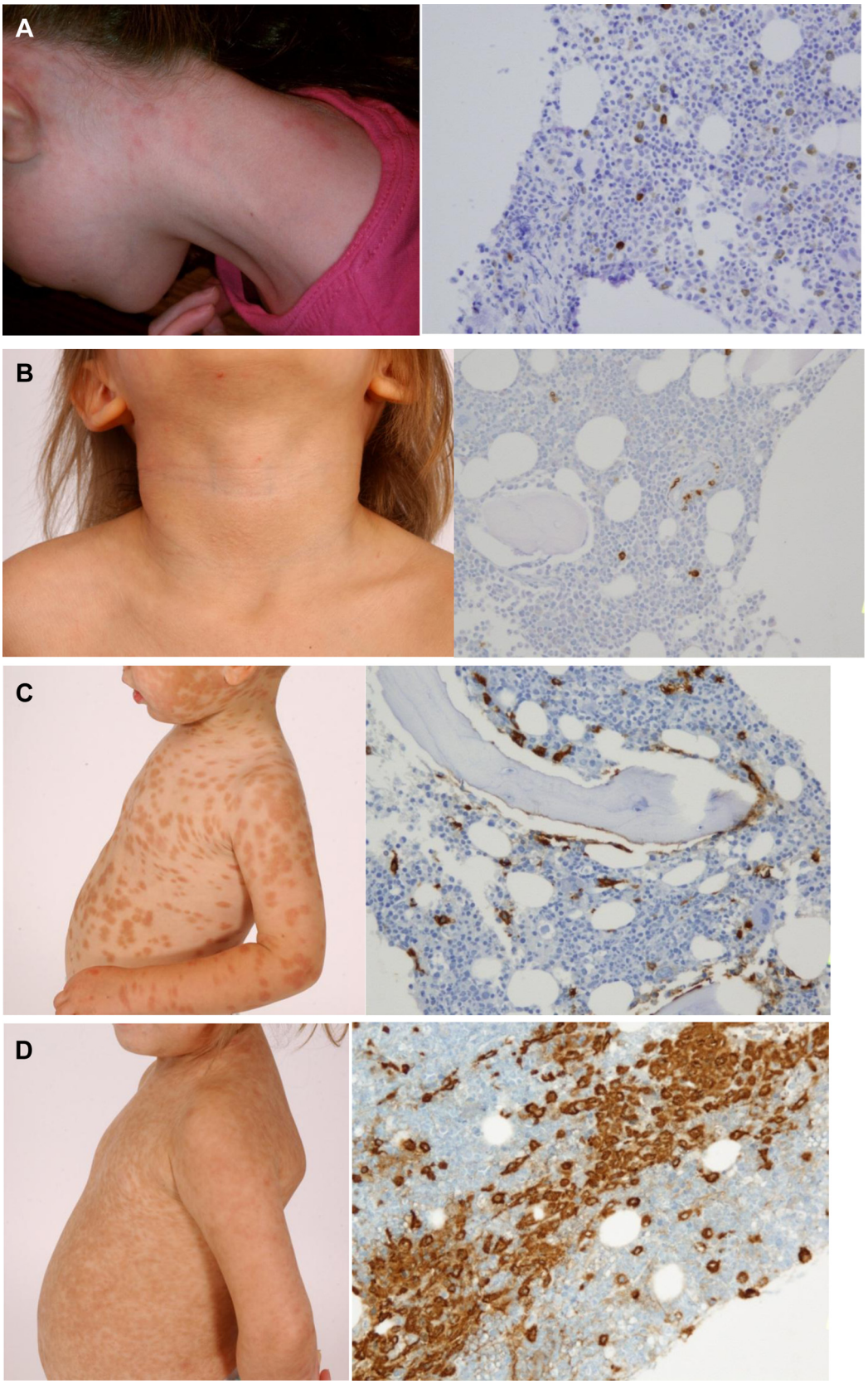

FIG E3. Skin appearance, bone marrow pathology, and serum tryptase levels in a patient with UP (tryptase level, 6.1 ng/mL; **A**), DCM (tryptase level, 26.0 ng/mL; **B**), and ISM (**C**: tryptase level, 53.7 ng/mL; **D**: tryptase level, 238.0 ng/mL). Immunohistochemistry staining for tryptase (dark brown) shows that greater than 25% of mast cells within the marrow were spindle shaped. All mast cells were $CD25^{+}$, and the D816V mutation was present. Magnification is ×200 for all bone marrow biopsy specimens.