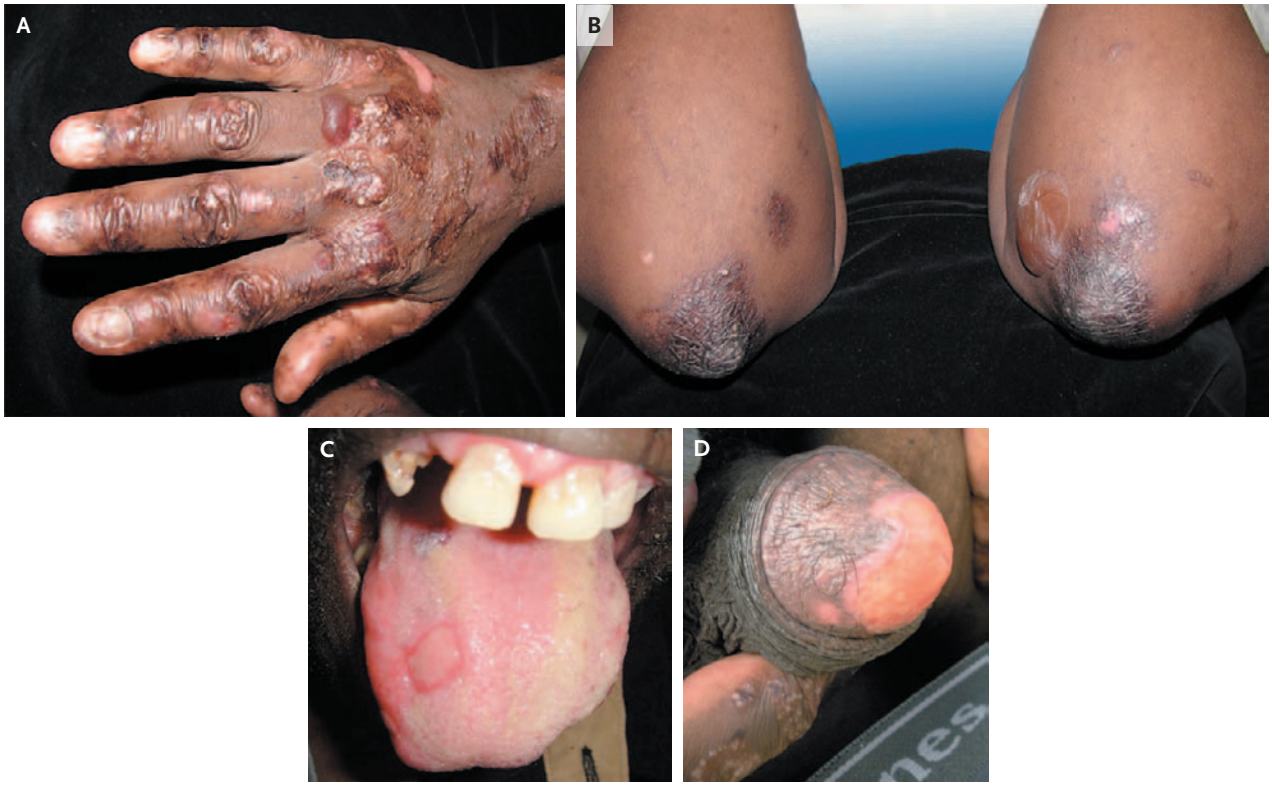
IMAGES IN CLINICAL MEDICINE

Epidermolysis Bullosa Acquisita



A 50-year-old man presented with tense noninflammatory bullae and erosions on trauma-prone areas of skin (Panels A and B) and the mucosae (Panels C and D). There was also atrophic scarring, milia formation, onychodystrophy, and anonychia. His medical history was remarkable for systemic lupus erythematosus (SLE), lupus nephritis, and hypertension. Examination of a skin-biopsy specimen revealed a cell-poor subepidermal vesicle. Direct immunofluorescence analysis revealed linear deposition of IgG at the dermal–epidermal junction (see the Supplementary Appendix). Studies of salt-split skin, which are used to examine the cutaneous basement-membrane zone for specific autoantibodies, supported the clinical suspicion of epidermolysis bullosa acquisita, an acquired subepidermal blistering disorder with autoantibodies against type VII collagen. This patient presented with the mechanobullous phenotype of epidermolysis bullosa acquisita, characterized by noninflammatory bullae that develop on trauma-prone acral skin and that heal with dyspigmentation, scarring, and milia formation. This autoimmune blistering disorder may mimic porphyria cutanea tarda, is often resistant to treatment, and may be associated with SLE. Treatment with systemic glucocorticoids, mycophenolate mofetil, and rituximab had limited benefit in this patient. Although he had a brief response to treatment with intravenous immune globulins, the administration of cyclophosphamide was started for maintenance of remission. The use of cyclophosphamide has resulted in the decreased formation of new blisters; however, disease activity has persisted.

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