



**Figure 25-34** Dermatitis herpetiformis. **A**, Lesions consist of intact and eroded (usually scratched) erythematous blisters, often grouped (seen here on elbows and arms). **B**, Selective deposition of IgA autoantibody at the tips of dermal papillae is characteristic. **C**, The blisters are associated with the accumulation of neutrophils (microabscesses) at the tips of dermal papillae. (**B**, Courtesy Dr. Victor G. Prieto, Houston, Texas.)

Porphyrias are pigments that are normally present in hemoglobin, myoglobin, and cytochromes. The classification of porphyrias is based on both clinical and biochemical features. The five major types are (1) congenital erythropoietic porphyria, (2) erythrohepatic protoporphyria, (3) acute intermittent porphyria, (4) porphyria cutanea tarda, and (5) mixed porphyria. Cutaneous manifestations consist of urticaria and vesicles associated with scarring that are exacerbated by exposure to sunlight. The vesicles are subepidermal in location and the adjacent dermis contains vessels with walls that are thickened by glassy deposits of serum proteins, including immunoglobulins (Fig. 25-36). The pathogenesis of these alterations is not understood.

## KEY CONCEPTS

### Blistering Disorders

- Blistering disorders are classified based on the level of epidermal separation.
- These disorders are often caused by autoantibodies specific for epithelial or basement membrane proteins that lead to unmooring of keratinocytes (acantholysis).
- Pemphigus is associated with IgG autoantibodies to various intercellular desmogleins, resulting in bullae that are either subcorneal (pemphigus foliaceus) or suprabasilar (pemphigus vulgaris).
- Bullous pemphigoid is associated with IgG autoantibodies to basement membrane proteins and produces a subepidermal blister.
- Dermatitis herpetiformis is associated with IgA autoantibodies to fibrils that bind the epidermal basement membrane to the dermis, and also produces subepidermal blisters.
- None inflammatory blistering disorders include inherited defects in proteins that stabilize the epidermis (e.g., epidermolysis bullosa) and inherited defects in porphyrin synthesis (the porphyrias) that lead to sun-induced skin damage through uncertain mechanisms.

## Disorders of Epidermal Appendages

### Acne Vulgaris

Virtually universal in the middle to late teenage years, acne vulgaris affects both males and females, although males tend to have more severe disease. Acne is seen in all races but is usually milder in people of Asian descent. It may be induced or exacerbated by drugs (corticosteroids, adrenocorticotropic hormone, testosterone, gonadotropins, contraceptives, trimethadione, iodides, and bromides), occupational exposures (cutting oils, chlorinated hydrocarbons, and coal tars), and conditions that favor occlusion of