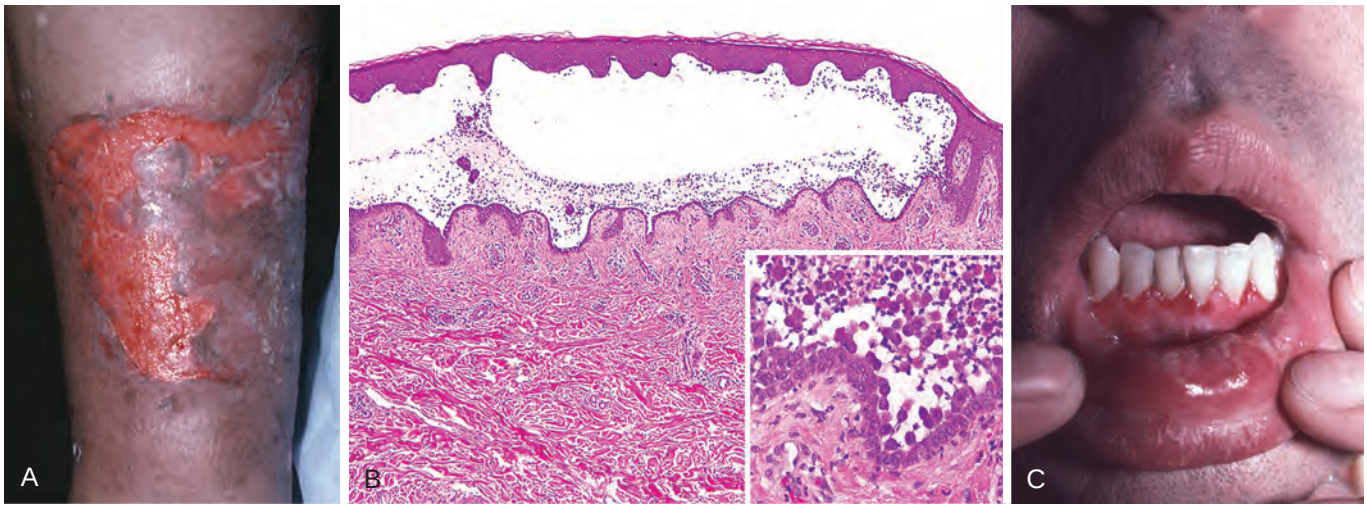

Figure 25-29 Pemphigus vulgaris. **A,** Eroded plaques are formed following the rupture of confluent, thin-roofed bullae, here affecting axillary skin. **B,** Suprabasal acantholysis results in an intraepidermal blister in which dyscohesive (acantholytic) epidermal cells are present *(inset)*. **C,** Ulcerated blisters in the oral mucosa are also common, as seen here on the lip.

- *Paraneoplastic pemphigus* occurs in association with various malignancies, most commonly non-Hodgkin lymphoma.

Pathogenesis. All forms of pemphigus are autoimmune diseases caused by IgG autoantibodies against desmogleins that disrupt intercellular adhesions and result in the formation of blisters. By direct immunofluorescence, lesions show a characteristic net-like pattern of intercellular IgG deposits. IgG is usually seen at all levels of the epithelium in pemphigus vulgaris, but tends to be more superficial in pemphigus foliaceus (Fig. 25-31). The distribution of desmoglein 1 and 3 in the epidermis and the presence of autoantibodies to one or both proteins appear to explain the position and severity of the blisters (Fig. 25-28). The antibodies cause these lesions primarily by disrupting the intercellular adhesive function of the desmosomes; they may also act indirectly by activating intercellular proteases. Paraneoplastic pemphigus arises most often in the setting of lymphoid neoplasms, and is also caused by autoantibodies that recognize desmogleins or other proteins involved in intercellular adhesion.

MORPHOLOGY

The common histologic denominator in all forms of pemphigus is **acantholysis**, the dissolution or lysis of the intercellular bridges that connect squamous epithelial cells. Acantholytic cells dissociate from one another, lose their polyhedral shape and become rounded. In pemphigus vulgaris and pemphigus vegetans, acantholysis selectively involves the cells immediately above the basal cell layer. In the vegetans variant, there is also overlying epidermal hyperplasia. An immediately **suprabasal acantholytic blister** is characteristic of pemphigus vulgaris (Fig. 25-29*B*). The single layer of intact basal cells that forms the blister base has been likened to a row of tombstones. In pemphigus foliaceus, blisters form by similar mechanisms but,

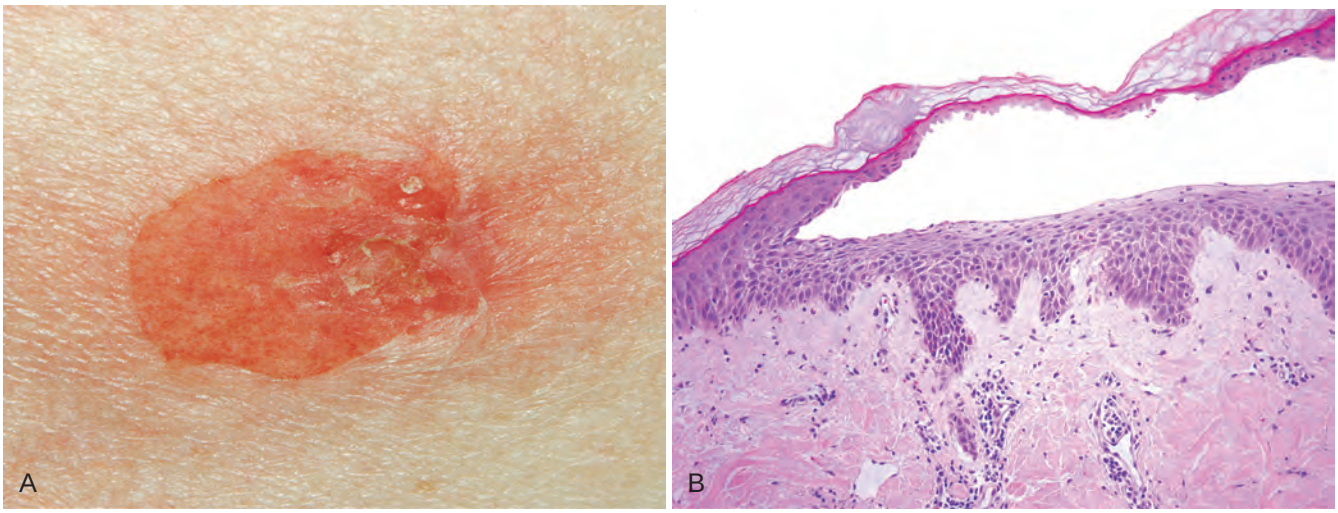

Figure 25-30 Pemphigus foliaceus. **A,** The delicate superficial (subcorneal) blisters are much less erosive than those seen in pemphigus vulgaris. **B,** Subcorneal separation of the epithelium is seen.