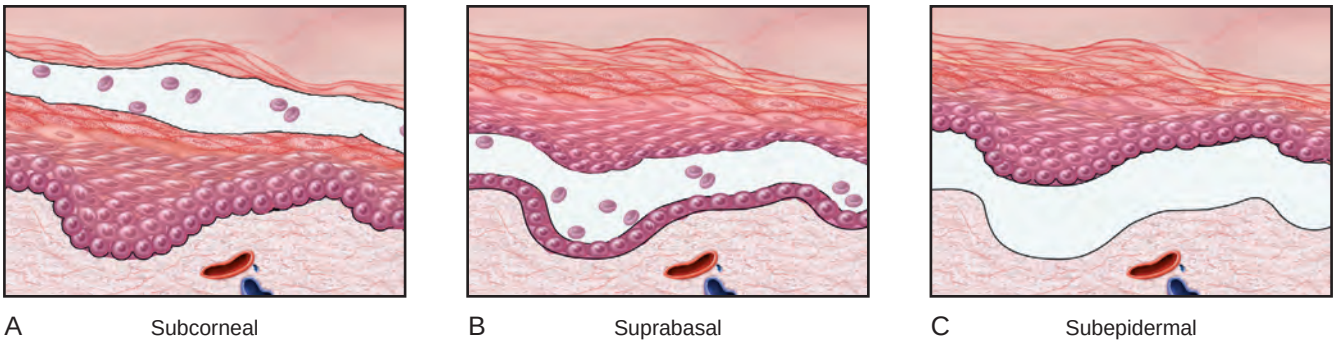

Figure 25-27 Schematic representation of different types of blisters. **A,** In subcorneal blisters, the stratum corneum forms the roof of the bulla (as in pemphigus foliaceus). **B,** In a suprabasilar blister, a portion of the epidermis, including the stratum corneum, forms the roof (as in pemphigus vulgaris). **C,** In a subepidermal blister, the entire epidermis separates from the dermis (as in bullous pemphigoid).

- *Pemphigus vulgaris*, by far the most common type (accounting for more than 80% of cases worldwide), involves the mucosa and skin, especially on the scalp, face, axilla, groin, trunk, and points of pressure. It may present as oral ulcers that may persist for months before skin involvement appears. Primary lesions are superficial vesicles and bullae that rupture easily, leaving shallow erosions covered with dried serum and crust (Fig. 25-29*A*).
- *Pemphigus vegetans* is a rare form that usually presents not with blisters but with large, moist, verrucous (wart-like), vegetating plaques studded with pustules on the groin, axillae, and flexural surfaces.
- *Pemphigus foliaceus* is a more benign form that is endemic in Brazil (where it is called *fogo selvagem*) and occurs sporadically in other geographic regions. Sites of predilection are the scalp, face, chest, and back, and the mucous membranes are only rarely affected. Bullae are so superficial that they mainly present as areas of erythema and crusting; these represent superficial erosions at sites of previous blister rupture (Fig. 25-30*A*).
- *Pemphigus erythematosus* is considered to be a localized, less severe form of pemphigus foliaceus that may selectively involve the malar area of the face in a lupus erythematosus-like fashion.

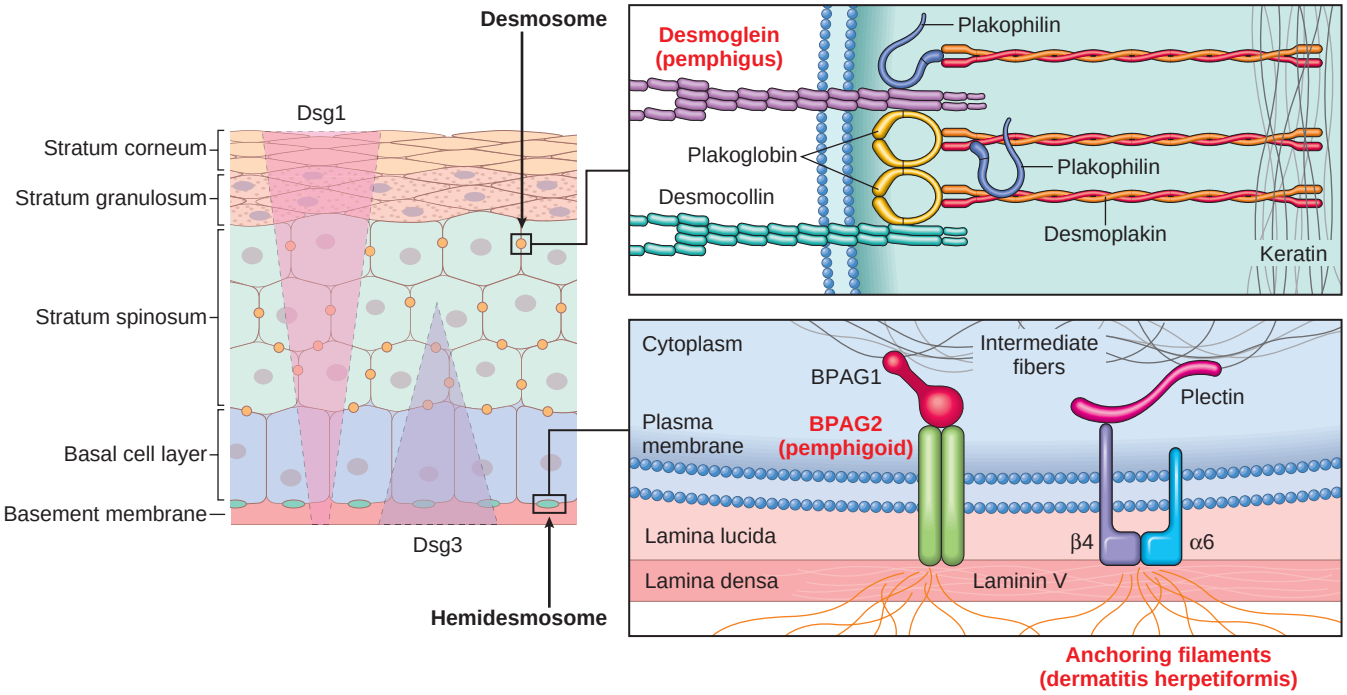

Figure 25-28 Keratinocyte adhesion molecules and blistering inflammatory disorders. Knowledge of the proteins composing desmosomes and hemidesmosomes is key to understanding blistering disorders. Desmogleins 1 and 3 (Dsg1, Dsg3) are functionally interchangeable components of desmosomes, but have different distributions within the epidermis *(left panel)*. The major structural proteins of desmosomes and hemidesmosomes are shown at *right*. In pemphigus vulgaris autoantibodies against Dsg1 and Dsg3 cause blisters in the deep suprabasal epidermis, whereas in pemphigus foliaceus the autoantibodies are against Dsg1 alone, leading to superficial, subcorneal blisters. In bullous pemphigoid autoantibodies bind BPAG2, a component of the hemidesmosomes, leading to blister formation at the level of the lamina lucida of the basement membrane. Dermatitis herpetiformis is caused by IgA autoantibodies to the fibrils that anchor hemidesmosomes to the dermis.