The best diagnosis is:

- a. Darier disease
- b. Hailey-Hailey disease
- c. herpesvirus infection
- d. pemphigus foliaceus
- e. pemphigus vulgaris
Hailey-Hailey disease (HHD), or benign familial chronic pemphigus, typically presents as suprabasal blisters with a perivascular and interstitial lymphocytic infiltrate (Figure 1). Villi, or elongated dermal papillae lined with a single layer of basal cells, protrude into the bullae (Figure 2). In HHD lesions, the epidermis is thickened with scale-crust, and at least the lower half of the epidermis shows acantholysis. Despite the acantholytic changes, a few intact intercellular bridges remain, giving the appearance of a dilapidated brick wall (Figure 2). There may be dyskeratotic cells among the acantholytic cells, though they are scant in many cases. These acantholytic dyskeratotic cells have eosinophilic polygonal-shaped cytoplasm. Hailey-Hailey disease typically does not show adnexal extension of the acantholysis. Direct immunofluorescence is negative in HHD.

Pemphigus vulgaris is an autoimmune intraepidermal bullous disease that presents with suprabasal acantholysis (Figure 3). The epidermis is not thickened and acantholysis is limited to the suprabasal layer. Acantholytic cells with eosinophils and/or neutrophils are found within the bullae. Perivascular and interstitial infiltrates of lymphocytes, eosinophils, and occasionally neutrophils are seen; however, the inflammatory cell infiltrate can vary from extensive to scant. Direct immunofluorescence usually reveals IgG and/or C3 deposition on the surface of the keratinocytes throughout the epidermis.
superficial perivascular and interstitial inflammatory cell infiltrate consisting of lymphocytes, eosinophils, and occasionally neutrophils. The amount of inflammatory cell infiltrate may vary. Bullous impetigo and staphylococcal scalded skin syndrome reveal a similar histopathologic pattern. Direct immunofluorescence usually discloses IgG and/or C3 deposition on cell surfaces of keratinocytes in the entire or upper epidermis.

Herpesvirus infection shows ballooning (intracellular edema) of keratinocytes. Eventually acantholysis occurs and intraepidermal bullae are formed. In the bullae, virus-associated acantholytic keratinocytes, some that are multinucleated, can be easily found (Figure 5). These cells are larger than normal keratinocytes and have steel gray nuclei with peripheral accentuation. Some of these cells are necrotic, and the remains of necrotic multinucleated acantholytic cells are easily recognized. Adnexal epithelial cells occasionally are affected by herpesvirus infection; nuclear change is similar to the epidermis. A perivascular and interstitial infiltrate of lymphocytes and neutrophils is seen. Neutrophils accumulate within the old bullae, clinically manifesting as a pustule.

Darier disease is characterized by suprabasal clefts and acantholysis above the basal layer (Figure 6). Similar to HHD, villi protrude within the clefts (Figure 6). Conspicuous columns of parakeratosis above the acantholytic epidermis often are observed. Dyskeratotic cells exist among acantholytic keratinocytes in the granular layer and parakeratotic column, which are known as corps ronds and crops grains, respectively. A scant to moderate lymphocytic infiltrate is found in the upper dermis.

REFERENCES