JOURNAL OF INSURANCE MEDICINE

J Insur Med 2004;36:267–268

GRAPHICS

Pemphigus

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Pemphigus vulgaris and the related pemphigus vegetans, pemphigus foliaceus and pemphigus erythematous, are associated with the highest mortality among primary skin diseases. Typical findings are illustrated.

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Key words: Mucous membranes, blisters, systemic infection, immunosuppressants.

Pemphigus is an acquired autoimmune disorder of the skin and mucous membranes most often affecting those between the ages of 30–70 years. The basic abnormality lies in the destruction of the mucopolysaccharide protein complex of intercellular cement. The tonofibrils within cells become disorganized and detached from desmosomes, followed by a dissolution of intercellular bridges in the epidermis just above the basal cell layer. This leads to a separation of epidermal cells so that bullae are formed within the epidermis, with a minimal amount of inflammation.

The disease in its more aggressive form commonly presents with mucous membrane lesions of the mouth, tongue and cheeks. Superficial ulcerations with peripheral extension are more often seen than intact blisters. The blisters are initially sparse, but soon become more generalized. The thin, flaccid, fluid-filled, superficial bullae quickly rupture to become painful, denuded, bleeding, weeping, non-healing erosions. Often times the normal skin between lesions can be removed simply by firm stroking.

The natural untreated course of this disease is slow progression with extensive denudation leading to fluid and electrolyte imbalance, metabolic derangements, sepsis, and death. Diagnosis is made by skin biopsy of early vesicles for routine histologic exam. Direct immunofluorescence shows deposits of immunoglobulins (usually IgG) and/or C3 in the intercellular spaces around keratinocytes. Antibodies to the intercellular areas of the epidermis may also be found in the serum of patients.

Pemphigus is classified by the level and type of splitting in the epidermis and by differences in the course of the disease. The more aggressive pemphigus vulgaris and its variant pemphigus vegetans are characterized by a suprabasilar split. The latter is so known because it heals with hypertrophic ''vegetative'' surfaces. Pemphigus foliaceus and its variant pemphigus erythematosus are characterized by a subcorneal split. These two more indolent forms of pemphigus tend to involve the scalp and face in a seborrheic dermatitis-like eruption, which often simulates the butterfly rash of systemic lupus er-

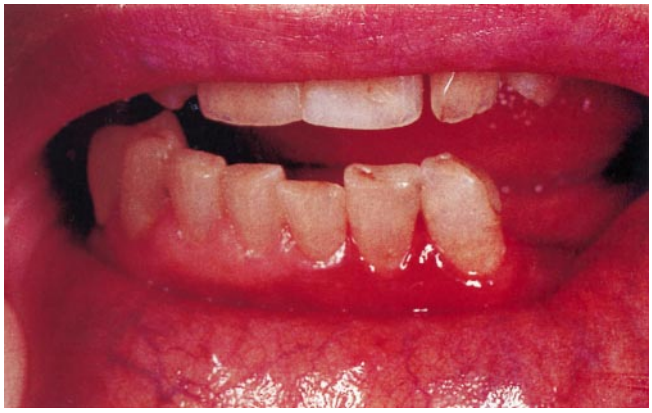

Figure 1. *Oral erosions usually precede the onset of skin blisters by weeks or months. From: Habif T, Campbell J Jr, Quitadamo M, Zug K.* Skin Disease Diagnosis and Treatment. *St. Louis, Mo; Mosby Inc: 2001. Reprinted with permission from Mosby an affiliate of Elsevier Science. ©2001 by Mosby, Inc.*

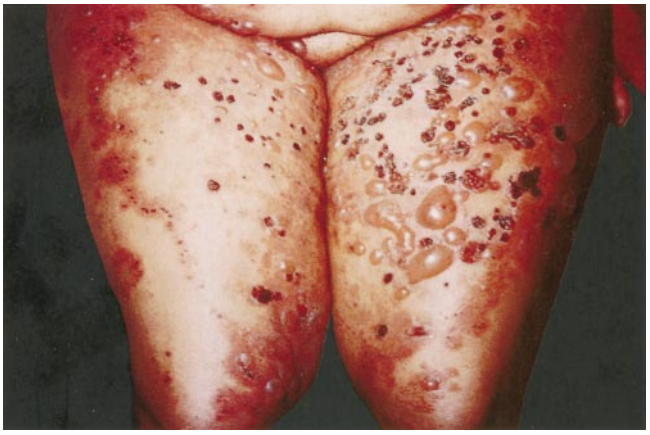

Figure 2. *Flaccid blisters rupture easily because the roof, which consists only of a thin portion of the upper epidermis, is very fragile. Healing is with brown pigmentation but without scarring. From: Habif T, Campbell J Jr, Quitadamo M, Zug K.* Skin Disease Diagnosis and Treatment. *St. Louis, Mo; Mosby Inc: 2001. Reprinted with permission from Mosby an affiliate of Elsevier Science. ©2001 by Mosby, Inc.*

ythematosis. Likewise, they seldom involve the mucous membranes. Lastly, familial benign pemphigus, or Hailey-Hailey disease, is a dominantly inherited disorder with a suprabasilar epidermal split. Involvement is more common in flexural areas (neck, axillae, groin), and may be precipitated by warm weather or superficial bacterial infections. It differs from other forms of pemphigus in its genetic pattern, absence of mouth lesions, benign course, and absence of intercellular antibodies.

Untreated, pemphigus vulgaris is invariably fatal in an average time of 14 months. Pemphigus vegetans runs a more prolonged course than pemphigus vulgaris, but early death without treatment remains the rule. In contrast, pemphigus foliaceus and erythematosus may follow a relatively benign course often lasting over a decade. High doses of systemic steroids (100–200 mg of Prednisone per day) over prolonged periods usually controls the disease. Methotrexate and other cytotoxic drugs are useful as steroid-sparing agents. Lower doses of medication will usually control the more indolent variants of pemphigus. Exacerbations of familial benign pemphigus may respond to systemic and/or topical antibiotics alone. The mortality of the more aggressive pemphigus variants remains high even with systemic steroid and immunosuppressive treatment. It may decrease over time once a remission is sustained.

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