Pemphigus Vulgaris
Pemphigus vulgaris develops primarily in older individuals, presenting with large and flaccid bullae. They...
Histopathology.
It is important that early blisters, preferably small ones, are selected for biopsy. Care should be given to keeping the blister intact with a finger (positive Asboe-Hansen sign). The newly created cleavage will reveal early and specific histologic changes.
The earliest recognized change may be either eosinophilic spongiosis rarely or, more commonly, "spongiosis" in the lower...
purpose, a smear is taken from the underside of the roof and from the base of an early, freshly opened bulla.
Because acantholytic keratinocytes are occasionally seen in various nonacantholytic vesiculobullous or pustular diseases, acantholysis, cytologic examination represents merely a preliminary test and should not supplant histologic examination.
Immunofluorescence (IF) Testing
The edge of a blister with intact surrounding normal skin, uninvolved skin adjacent...
remission may be a good prognostic indicator. At this time, DIF testing is incorrectly thought by most to
of false positives; however, false positives may occur. On occasion, it may be difficult to distinguish intercellular
staining of pemphigus from nonspecific staining; for example, spongiotic dermatitis, psoriasis, bullous impetigo.
and epidermis adjacent to ulcers secondary to a number of disorders may have squamous intercellular substance
IgG due to the presence of serum in the intercellular substance. Often, IgM, IgA, fibrinogen, and albumin...
present as well, indicating nonspecific trapping in the false-positive tests. Immunoperoxidase methods have
achieved roughly the same sensitivity as the IF method, but they have not replaced IF testing as the prime
diagnostic tool. For IIF testing, unfixed frozen sections of guinea pig esophagus, monkey esophagus, or
human skin are used as substrate. In general, monkey esophagus is the best substrate for IIF tests. Circu
IgG autoantibody is demonstrated in the squamous intercellular substance in 80% to 90% of cases, and...
titer correlates with disease activity. False-positive IIF tests occur. In a series of 1,500 patients with circulating pemphigus antibodies, approximately 1% had no evidence of clinical disease.
vitro deposition in stratified squamous epithelium in the absence of pemphigus have been reported in burns,
penicillin allergy, toxic epidermal necrolysis (TEN), systemic lupus erythematosus (SLE), myasthenia gravis
bulla pemphigoid, cicatricial pemphigoid, and lichen planus and in patients with antibodies directed ag
blood groups A and B. Such antibodies are present in low titer and are thought to be nonpathogenic.
Antidesmoglein autoantibodies are sometimes found in patients with no bullous disease. For example, the...
been found in patients with silicosis and in relatives of patients with pemphigus vulgaris.
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**Pathogenesis**

Compelling evidence has accumulated that IgG autoantibodies against desmoglein 3 and desmoglein 1 are pathogenic and disrupt the normal function of these proteins.
are thought to play an important role in acantholysis. Although complement fixation by pemphigus antibodies
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promote acantholysis, acantholysis occurs in experimental systems in the absence of complement. The stimulus for the...

**Ultrastructural Study**

The intercellular cement substance, or glycocalyx, is partially or entirely lysed in lesions with early acantholysis. As acantholysis progresses, the desmosomes gradually disappear and the keratinocytes develop numerous cytoplasmic...
processes that often interdigitate with one another. All of the early ultrastructural changes in pemphigus
In early blisters that are free of secondary changes, such as the degeneration or regeneration of epidermal cells, the gold standard in diagnosis at this time and must be pursued in all cases in which pemphigus vulgaris is considered.

*Differential Diagnosis*
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