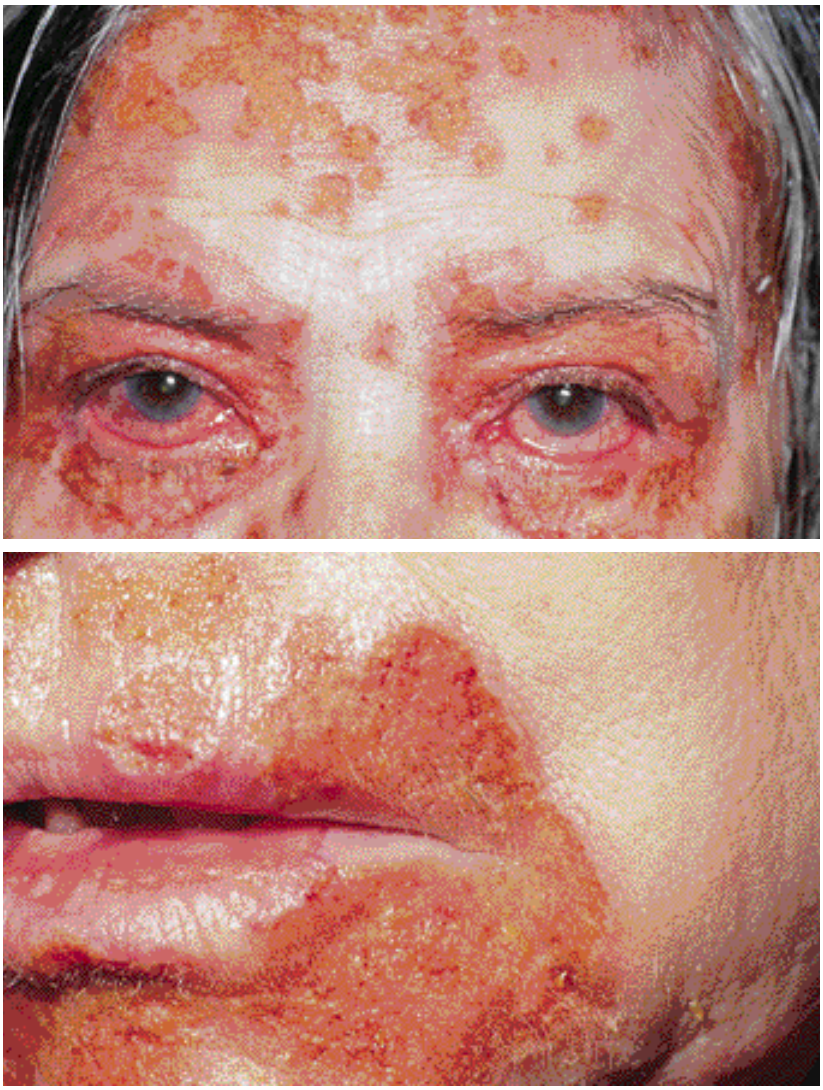
What Is Your Diagnosis?



A patient with pemphigus vulgaris presented with worsening of her erosions.

PLEASE TURN TO PAGE 317 FOR DISCUSSION

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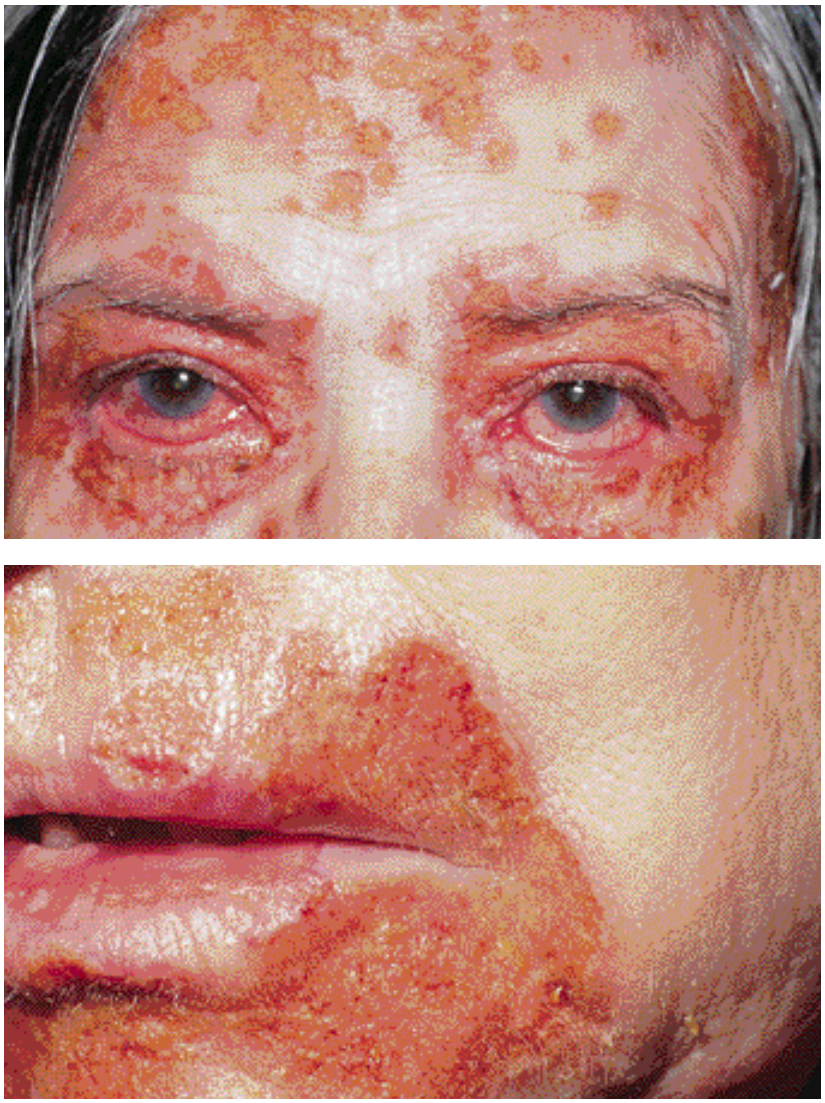
The Diagnosis: Eczema Herpeticum Superinfection in a Patient With Pemphigus Vulgaris



Eczema herpeticum is traditionally termed *Kaposi's varicelliform eruption*, referring to any generalized eruption secondary to a virus in a patient with skin disease. Because most of the associated eruptions are caused by infection with herpes simplex viruses (HSV), the preferred term is *eczema herpeticum*. Eczema herpeticum is a widespread infection with HSV (either HSV-1 or HSV-2), which generally occurs in a patient with preexisting skin disease.[1] Examples of preexisting skin disease include: atopic dermatitis, benign familial pemphigoid, bullous pemphigoid, Darier disease, icthyosiform erythroderma, icthyosis vulgaris, impetigo, mycosis fungoides, pemphigus foliaceus, pemphigus vulgaris, scabies, seborrheic dermatitis, and Sézary syndrome.[2] Epidermal barrier dysfunction and immunosuppressive therapy are predisposing factors for superinfection, as evidenced in the case presented.

The diagnosis of eczema herpeticum may be masked by the primary disease. Herpes superinfection of the geographic erosions of pemphigus vulgaris

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may be interpreted as acute worsening of the patient's primary disease. There are multiple reports in the literature of "worsening pemphigus" later diagnosed as herpetic superinfection because the herpetic infection typically involves only previously eroded skin.[3] Distinction between the erosions of pemphigus and herpes superinfection may be made by noting the more "punched-out," circular, often crusted erosions resulting from HSV infection. Small grouped vesicles are often visible at the edge of pemphigus erosions.[4]

Other variables aid in diagnosing eczema herpeticum in the setting of pemphigus. Onset of fever associated with a "pemphigus" flare should immediately spark investigation for herpetic infection.[5] Another key diagnostic variable is burning pain beyond the discomfort normally associated with the primary disease.[6] As seen in our patient, involvement of tissues that are relatively low in pemphigus antigens (such as ocular tissue) suggests a disease process other than pemphigus.[7] A negative medical history for herpetic infection should not preclude the diagnosis of eczema herpeticum.[6,8] The prevalence of HSV-1 in the general population worldwide is as high as 85%.[9]

Tzanck test results can confirm the diagnosis of eczema herpeticum. Multinucleated giant cells with multiple nuclei molded together in a formation resembling a jigsaw puzzle are suggestive of herpetic infection.[9] Results of direct florescent antibody testing and HSV serotyping (often available within several hours of presentation) allow for rapid confirmation of the clinical diagnosis. Viral culture results play a confirmatory role in the diagnosis because they are not available for at least 2 days after the culture is taken. Physicians should consider taking viral cultures from other sites (eg, eye, oropharynx) if the patient has associated complaints. Hematoxylin and eosin stain of affected skin tissue reveals loss of epidermis, ballooning necrosis of keratinocytes, and multinucleated giant cells.[8]

Herpetic infection in an immunosuppressed patient does not always remain localized to the skin. There are reports of herpes keratitis in patients with pemphigus.[7] Symptoms of keratitis include blurring of vision, pain, and swelling of the eyelids. Examination of the affected eye will reveal dendritic keratitis. Fatal cases of hepatic involvement and disseminated intravascular coagulation have been reported in patients with pemphigus.[8]

Acyclovir is the most widely used treatment for eczema herpeticum. This drug is phosphorylated by HSV viral thymidine kinase then phosphorylated twice more in the infected host cell before it is incorporated into DNA, acting as a chain terminator.[1] For eczema herpeticum, the usual dose is 15 mg/kg per day of acyclovir for a duration of at least 5 days. This may be administered either orally or intravenously, depending on the patient's ability to take oral medications. Evidence of other organ involvement requires prolonged treatment.[6] Published case reports suggest a need for continuing antiviral prophylaxis if the patient suffers recurrent herpetic erosions.[4] Adequate pain control is also central to treatment of acute eczema herpeticum.

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