BULLOUS SKIN DISEASES

Skin and Mucous Membrane Blisters in Autoimmune Disease

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The autoimmune bullous skin disorders may occur alone, for instance in patients with pemphigus, or in people with other autoimmune disorders, such as systemic lupus erythematosus (SLE). Bullous skin disorders may also occasionally occur in pregnancy.

Symptoms

Bullous skin diseases are characterized by the presence of blisters or erosions of the skin and mucous membranes. In some bullous disorders such as pemphigus vulgaris, blistering is the primary disease manifestation. In other disorders, such as lichen planus or systemic lupus erythematosus (SLE), blisters occur infrequently. Bullous SLE occurs in less than 5 percent of patients, and some studies suggest that there is an increased incidence of the bullous variant of SLE in young black women.

Acquired, Induced, or Autoimmune

Bullous skin diseases may be acquired or induced or they may be autoimmune in origin. The autoimmune bullous skin disorders are all characterized by the presence of autoantibodies that target distinct adhesion molecules of the epidermis and dermoeipidermal basement membrane zone. The consequences of these antibodies are a loss of the targeted protein's adhesive properties, which leads, in turn, to the appearance of blisters and erosions. Like most autoimmune disorders, autoimmune bullous or vesiculobullous skin disorders are more likely to occur in women. Women of childbearing age have the highest risk for developing autoimmune bullous diseases.

The autoimmune bullous disorders include:

• Pemphigus and Pemphigoid disorders

• Epidermolysis bullosa acquisita

• Dermatitis herpetiformis

• Herpes (pemphigoid) gestationis

• IGA - mediated disorders

Pemphigus and Pemphigoid Disorders
Pemphigus disorders and pemphigoid disorders (bullous pemphigoid, cicatrical pemphigoid) are considered prototypic bullous disorders because of their well-defined autoantibody-mediated development or pathogenesis. The three pemphigus disorders target desmoglein protein, and the pemphigoid disorders target Type XVII collagen and bullous pemphigoid (BP) antigen 2, BP antigen 180, BP antigen 230, and laminin 5. The end result is a loss of the skin's architecture. In the absence of a well-organized viable skin composition, blisters and erosions form craters between the tissues cells.

Pemphigus disorders may present or worsen during pregnancy, especially in the first and second trimesters. Menstruation is reported to trigger or cause relapses of pemphigus. Other suspected risk factors for pemphigus include pregnancy, frequent handling of certain spices, repeated minor thermal burns, and cosmetic procedures.

Bullous pemphigoid primarily affects the skin, especially the lower abdomen, groin, and flexor surfaces of the hands and feet. About 10-40 percent of patients have mucous membrane involvement. The common age of onset is 65 to 75 years.

Cicatricial pemphigoid (CP) usually targets people between the ages of 60-80 although it has been reported in people of all ages, including children. Several subgroups of CP have been described including antiepiligrin CP, Pure Ocular CP, which affects only the eyes, Anti-BP Antigen Mucosal Pemphigoid, and Oral Pemphigoid.

**Dermatitis Herpetiformis and IgA-mediated Autoimmune Disorders**

Dermatitis herpetiformis is a bullous skin disorder that occurs in patients with gluten sensitivity. It may also occur in patients with autoimmune thyroid disease, type 1 diabetes, vitiligo, and rheumatoid arthritis. The blisters usually appear on the buttocks and back and are known to cause severe itching.

Blister in dermatitis herpetiformis are caused by reduced deposits of immunoglobulin A (IgA) and complement, and also autoantibodies to reticulin, gliadin, and gluten. Lasting remission occurs with continued use of a gluten-free diet. A similar condition of IgA mediated dermatosis that does not involve gluten sensitivity is also mediated by IgA and causes "sausage-like" lobulated blisters arranged at the outer edges of flattened erythematous patches.

**Herpes and Pemphigoid Gestationis**

Herpes gestationis (HG) is seen in pregnancy, molar pregnancy, trophoblastic malignancy, hormonal perturbation in susceptible women, and during the postpartum period. It's characterized by gyrate erythema (skin redness) and blisters. Herpes gestationis often worsens just before, during, or after delivery, and it can recur in subsequent pregnancies. Rarely, the skin of newborns born to mothers with HG can be transiently affected due to passive transfer of antibodies. Association with other autoimmune diseases, especially Graves' disease, has been reported in 11 percent of
patients with HG in one study of 75 patients. Dr. Bernard Bihari has been treating herpes gestationis with low dose naltrexone (LDN) successfully for more than 20 years.

**Epidermolysis Bullosa Acquista**

Epidermolysis bullosa acquisita is a chronic, uncommon, subepidermal blistering disease of the skin and mucous membranes. The average age of onset is 50 years although people of all ages, including children, may be affected.

**Treatment**

Treatment of autoimmune bullous disease with topical corticosteroids has little value unless the blisters are localized to only one area. Most patients with bullous disorders are treated with oral doses of corticosteroids and other immunosuppressive drugs such as azathioprine or cyclophosphamide. Patients with dermatitis herpetiformis are also treated with dapsone unless they are allergic to sulfonamides. All of these oral treatments have the potential for causing severe side effects. Patients on these medications must have frequent blood tests to monitor these effects. Other corticosteroid-sparing drugs used as therapies include levamisole and chloroquine. In the early stages of bullous pemphigoid treatment with topical corticosteroids and oral tetracycline has been found to be effective, causing remission in 1-3 weeks.

**Resources:**


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