Overview of Bullous Pemphigoid

Bullous pemphigoid is an autoimmune disease, that usually attacks the elderly patient. It occurs when the body’s defense mechanism produces autoantibodies that interact with the bullous pemphigoid antigen [BPAG1 and BPAG2] in hemidesmosomes of basal keratinocytes (that join the keratocytes to the basement membrane zone), which is followed by complement activation and recruitment of inflammatory cells such as neutrophils & eosinophils. This results in characteristic skin blisters.

While Bullous pemphigoid typically affects the skin, it may also involve the eyes, mouth and genital area. Even though the condition develops insidiously in the majority of the cases, there are some drugs that trigger the development of bullous pemphigoid. Bullous pemphigoid is found more in people older than 60 years, but it can also develop in younger persons. This disease is not curable, but the treatment aims to reduce the symptoms.

Symptoms

The condition is characterized by generalized eruption with confluent urticarial plaques
and multiple tense blisters. The condition is severely pruritic.

**Description of the lesion:**

Erythematous, pruritic papular or urticarial-type lesions that may precede the formation of the bullae by months. It is characterized by tense blisters, as opposed to the flaccid blisters in pemphigus. The eruption may be localized or generalized around the body. Oral lesions are very rare. At times, these blisters break, leaving an exposed skin that is open and painful.

**Diagnosis**

A thorough history should be taken to enable the physician to rule out other conditions. The skin should be assessed, blood sample and skin biopsies carried out.

**Investigations**

- Antibasement membrane IgG autoantibodies can be found in the serum.
- Immunofluorescence microscopy reveals:

**Treatment**

The main aim of therapy is to reduce itching and blisters and assist the proper skin healing. Usually, the choice of treatment depends on the extent of skin involvement.

**Immunosuppressive agents**

- In very mild cases and for local recurrences, topical glucocorticoid or topical tacrolimus
- Systemic prednisone in severe cases, either alone or combined with azathioprine
- Low-dose MTX is effective & safe in elderly

**Overview of Pemphigus Vulgaris**

Pemphigus Vulgaris is a rare autoimmune condition that results in blisters on the skin and mucous membranes (mouth, throat, nose, eyes, genitals, lungs), which are usually painful.

This condition occurs due to loss of the normal cell-to-cell adhesion in the epidermis (*acantholysis*) as a result of circulating Auto-Antibodies of the IgG class; which bind to desmosomes (desmogleins 3) that join the keratocytes in the epidermis.

Pemphigus Vulgaris is the most prevalent kind of a cluster of autoimmune disorders called pemphigus. Respective kinds of pemphigus are distinguished by the location of the blisters. The condition begins with a blister on the mouth, which then goes to the skin and at times the genitals. If left untreated, pemphigus Vulgaris can be life-threatening.

Pemphigus Vulgaris is mainly genetic, and affect individuals of all races, genders, and ages. However, it is more prevalent in individuals from the Mediterranean areas, Eastern European Jews, people residing in the Amazon of Brazil and middle-aged and old people.
Symptoms

The main symptoms of pemphigus Vulgaris are painful blisters that usually starts in the oral mucosa (rapidly become erosive & painful preventing eating), and months may elapse before skin lesions occur.

Lesions might be just localized for several months, after that generalized bullae may occur.

**Description of the lesion:**

It is characterized by flaccid bullae (blisters) that appear spontaneously, after which may rupture. When these blisters rupture, they become *tender and painful from erosions.*

These extensive erosions can bleed easily.

Diagnosis

Positive Nikolsky’s sign: It’s easy separation of the epidermis on superficial pressure, and this sign is considered a characteristic for Pemphigus Vulgaris.

A positive Nikolsky’s sign occurs if the patient skin peels away easily when the surface is wiped with a ball of cotton wool. It may also be necessary to carry out a biopsy of the skin to confirm that it is pemphigus Vulgaris.

Investigations

1. Immunofluorescence microscopy reveals *IgG deposits intercellularly in the epidermis*
2. IgG Autoantibodies to the intercellular substance of epidermis (IIF).
3. ELISA: Antibodies to desmoglein 3 more than desmoglein 1.

Treatment

The most effective and important treatment for this disease is corticosteroids. High doses are needed initially, which can then be lowered as the blisters reduce.

Additional drugs such as azathioprine, methotrexate that can reduce the activities of the immune system can also be used with prednisone.

Using Immunofluorescence to Differentiate Between Bullous Pemphigoid and Memphis Vulgaris

The direct immunofluorescence examination (DIF) is usually best to utilize while exploring skin lesions, depending on the disease being investigated. The biopsy should be at least 4 mm deep so that the epidermis and dermis can be gathered equally. Furthermore, the biopsy sample will produce better results if it undergoes and little trauma as possible. The most recent lesions should be used while taking a sample for biopsy.
Direct immunofluorescence examination

**Pemphigus Vulgaris**

Patients with pemphigus Vulgaris present with IgG autoantibodies against desmosomes, showing that the prognosis is not too good.

Direct immunofluorescence examination shows that there are intercellular deposits (net-like band) of IgG and C3 in the epidermis.

If the patients’ mucous membrane is involved and the disease is still active, the C3 is mostly found in the lower parts of the epithelia.

**Bullous pemphigoid**

Direct immunofluorescence examination in all patients with bullous pemphigoid reveals linear collections along the hemidesmosomes (dermal-epidermal junction) with anti-C3 conjugate and IgG.

It is very rare to see IgA and IgM with DIF.

These biopsies are usually taken around the joints, where a high quantity of BP antigen is found.

**References**


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