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BULLOUS PEMPHIGOID IS THE MOST COMMON AUTOIMMUNE SUBEPIDERMAL BLISTERING DISEASE IN GERIATRIC PERSONS

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ABSTRACT:

Pemphigus is a disease that causes blistering of the skin and the inside of the mouth, nose, throat, eyes, and genitals. The disease is rare in the United States. Pemphigus is an autoimmune disease in which the immune system mistakenly attacks cells in the top layer of the skin (epidermis) and the mucous membranes.Pemphigus vulgaris is caused by a problem with the immune system, which is the body's defense against infection. Instead of attacking germs, something goes wrong with the immune system and it attacks the skin by mistake. This damages the skin and causes blisters to appear.Bullous pemphigoid is a rare skin condition that mainly affects older people. It usually starts with an itchy, raised rash. As the condition develops, large blisters can form on the skin.It may last a few years and sometimes causes serious problems, but treatment can help manage the condition in most cases.

KEYWORDS: Skin, Epidermis, Dermis, Itching, Pemphigus, Bullous, ELISA.

OVERVIEW

Bullous pemphigoid is a rare skin condition that mainly affects older people. It usually starts with an itchy, raised rash. As the condition develops, large blisters can form on the skin. It may last a few years and sometimes causes serious problems, but treatment can help manage

the condition in most cases. Topical corticosteroids, systemic corticosteroids, and doxycycline are the mainstays of initial treatment for bullous pemphigoid. Additional immunomodulatory therapies are often added to minimize the adverse effects of chronic corticosteroid therapy or to augment improvement in the disease. The most common treatment is prednisone, which comes in pill form. But long-term use can increase your risk of weak bones, diabetes, high blood pressure, high cholesterol and infection. Corticosteroid ointment can be rubbed on your affected skin and causes fewer side effects.[1]

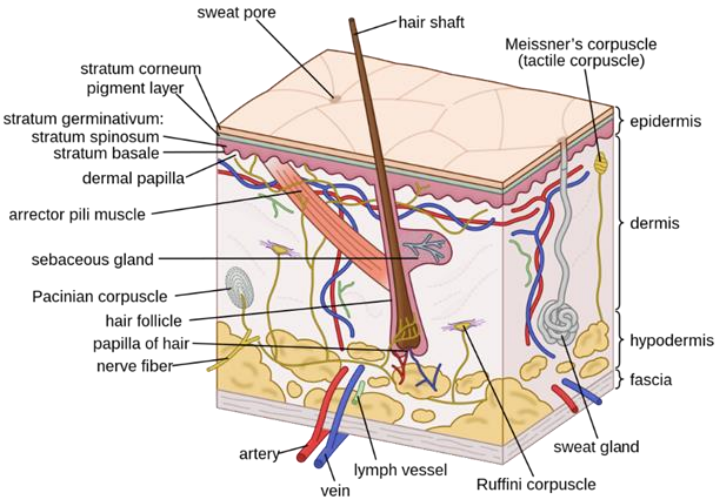

Figure-1: Skin anatomy.

Bullous pemphigoid is a chronic autoimmune skin disorder resulting in generalized, pruritic, bullous lesions in older patients. Mucous membrane involvement is rare. Diagnosis is by skin biopsy and immunofluorescence testing of skin and serum. Topical and systemic corticosteroids are used initially. Bullous pemphigoid (BP) is the most common autoimmune subepidermal blistering disease, affecting predominantly elderly people. It is an autoimmune disorder that occurs when the body's immune system attacks and destroys healthy body tissue by mistake. Specifically, the immune system attacks the proteins that attach the top layer of skin (epidermis) to the bottom layer of skin.It eventually goes away on its own, but it can last a few years.[2] Treatment can help your skin heal, stop new patches or blisters appearing, and reduce the chance of your skin getting infected. Internal medications: Medications like *Guluchyadi kasayam, Mustadi kasaya, Kulakadi kasaya, Chandanasava*, etc. are prescribed in order to balance the vitiated *doshas* and help in a bullous pemphigoid condition. Other medications are also prescribed to treat bullous pemphigoid. Tetracycline, doxycycline, and dapsone are antibiotics, which can reduce the inflammation inside your body. For some

patients, applying a corticosteroid cream or ointment and taking an antibiotic provides effective treatment. Among all the treatment modalities in India, the most commonly used is steroids, either oral or in the form of pulse therapy. Non-steroidal immunosuppressive drugs are added as adjuvants to increase the efficacy and to have a steroid sparing effect. No, bullous pemphigoid isn't contagious. You can't spread bullous pemphigoid to another person through skin-to-skin contact. Oral terbinafine has been associated with the development of bullous pemphigoid. Vancomycin is the most common cause of drug-induced LAD[left anterior descending artery]. Other drugs known to cause LAD include diclofenac, somatostatin, lithium, phenytoin, captopril, amiodarone, cefamandole, amoxicillin, and ampicillin-sulbactam.[3]

Foods that patients have reported to be bothersome (you may want to talk with your doctor about avoiding these):Citrus, Acidic Fruits, Bagels, Garlic, Potato Chips, Barbeque/cocktail sauces, Horseradish, Relishes. However, no increased overall cancer risk was found in patients with bullous pemphigoid. It is the most common autoimmune blistering disease.

Detection: Enzyme-linked immunosorbent assay. The ELISA technique analyzes the bullous pemphigoid antigen-specific IgG autoantibodies in the patients' sera by using various lengths of recombinant proteins of the BPAg1 or BPAg2 antigens. In several reports, ELISA has been demonstrated to be highly sensitive and specific.[4]

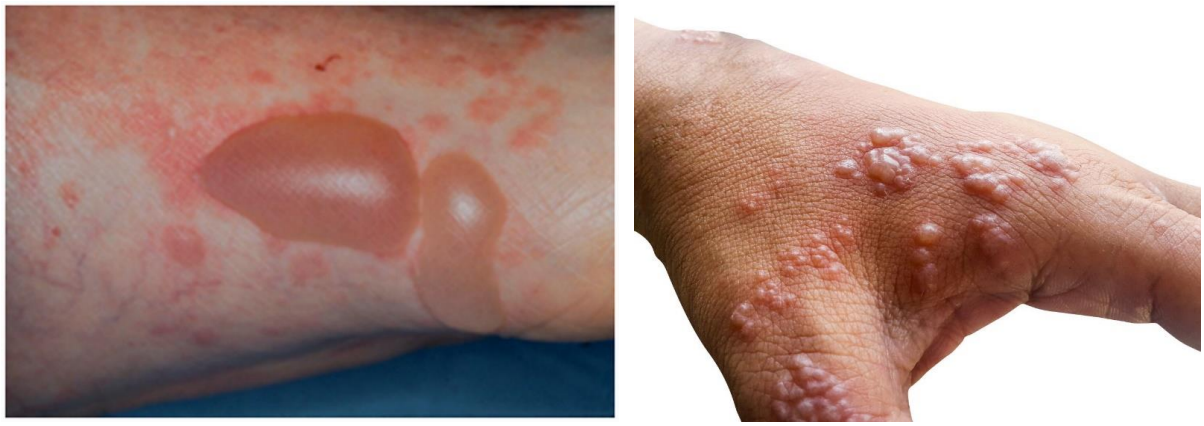

Figure-2: Bullous pemphigoid.

Precautions from Bullous pemphigoid: Protect your skin from irritation and injury. Bullous pemphigoid makes skin fragile. Wash your hands, Look for signs of infection every day, Follow your treatment plan, Care for mouth sores, Keep all follow-up medical appointments, Get support.

Bullous pemphigoid can last from 1 to 5 years. If untreated, the blisters and open areas of your skin can cause pain and discomfort. You may also get a severe infection in the open areas of your skin.

Bacterial staphylococcal and streptococcal skin infection, and sepsis.
Viral infection with herpes simplex, varicella or herpes zoster.
Complications of treatment.
Underlying and associated diseases.

It commonly affects people older than 60 years but can occur in younger people. Once someone is diagnosed as having this disease, they can have it for many years. Treatment helps to control the disease, but there is no permanent cure. Severe itching and blisters occur in almost all patients. It often goes away on its own in a few months, but may take as many as five years to resolve. Treatment usually helps heal the blisters and ease any itching. It may include corticosteroid medications, such as prednisone, and other drugs that suppress the immune system.The median age of death was 89.6 years and the mean was 88.5 (SD 7.55).[5]

Vitamin D: One of the most important vitamins needed for Pemphigus and Pemphigoid patients as the combination of lack of sun, the use of Prednisone and even the disease itself can deplete the body's absorption of this vitamin. Bullous pemphigoid is an autoimmune disease. This type of disease develops when your immune system malfunctions, causing your body to attack a part of itself.Bullous pemphigoid (a type of pemphigoid) is an autoimmune pruritic skin disease which typically occurs in people aged over 60, that may involve the formation of blisters (bullae) in the space between the epidermal and dermal skin layers. It is classified as a type II hypersensitivity reaction, which involves formation of anti-hemidesmosome antibodies, causing a loss of keratinocytes to basement membrane adhesion.

Clinically, the earliest lesions may appear as a hives-like red raised rash, but could also appear dermatitic, targetoid, lichenoid, nodular, or even without a rash (essential pruritus). Tense bullae eventually erupt, most commonly at the inner thighs and upper arms, but the trunk and extremities are frequently both involved. Any part of the skin surface can be involved. Oral lesions are present in a minority of cases. The disease may be acute, but can last from months to years with periods of exacerbation and remission. Several other skin diseases may have similar symptoms. However, milia are more common with epidermolysis bullosa acquisita, because of the deeper antigenic targets. A more ring-like configuration with

a central depression or centrally collapsed bullae may indicate linear IgA disease. Nikolsky's sign is negative, unlike pemphigus vulgaris, where it is positive.In most cases of bullous pemphigoid, no clear precipitating factors are identified. Potential precipitating events that have been reported include exposure to ultraviolet light and radiation therapy. Onset of pemphigoid has also been associated with certain drugs, including furosemide, nonsteroidal anti-inflammatory agents, DPP-4 inhibitors, captopril, penicillamine, and antibiotic.[6]

Drug of choice: Glucocorticoids [clobetasol & halobetasol]. Glucocorticoids are the mainstay of treatment for most bullous disorders. Azathioprine. Azathioprine is a purine analog used as a steroid-sparing agent for autoimmune bullous diseases. Mycophenolate mofetil. Cyclophosphamide. Methotrexate. Cyclosporine.Dapsone. Tetracycline and Niacinamide.

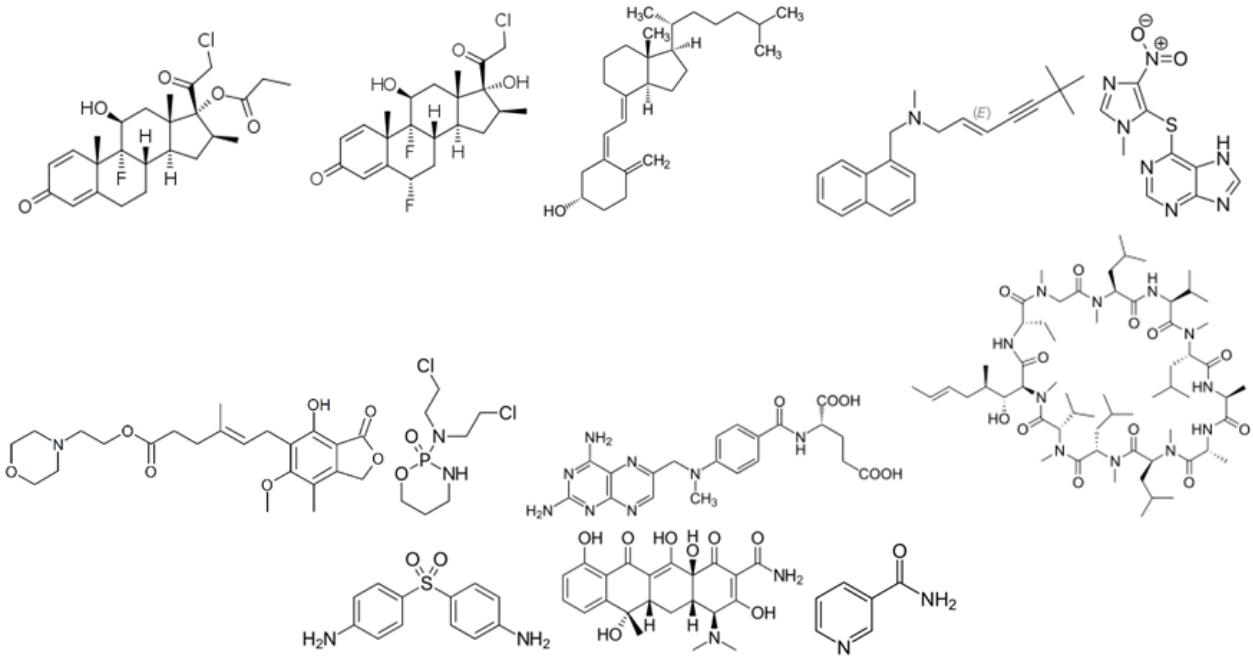

Figure-3: Drug of choice.

The bullae are formed by an immune reaction, initiated by the formation of IgG autoantibodies targeting dystonin, also called bullous pemphigoid antigen and/or type XVII collagen, also called bullous pemphigoid antigen which is a component of hemidesmosomes. A different form of dystonin is associated with neuropathy. Following antibody targeting, a cascade of immunomodulators results in a variable surge of immune cells, including neutrophils, lymphocytes and eosinophils coming to the affected area. Unclear events subsequently result in a separation along the dermoepidermal junction and eventually stretch

bullae.Treatments include topical steroids such as clobetasol, and halobetasol which in some studies have proven to be equally effective as systemic, or pill, therapy and somewhat safer. However, in difficult-to-manage or widespread cases, systemic prednisone and powerful steroid-free immunosuppressant medications, such as methotrexate, azathioprine or mycophenolate mofetil, may be appropriate. Some of these medications have the potential for severe adverse effects such as kidney and liver damage, increased susceptibility to infections, and bone marrow suppression. Antibiotics such as tetracycline or erythromycin may also control the disease, particularly in patients who cannot use corticosteroids.[7]

The anti-CD20 monoclonal antibody rituximab has been found to be effective in treating some otherwise refractory cases of pemphigoid. A 2010 meta-analysis of 10 randomized controlled trials showed that oral steroids and potent topical steroids are effective treatments, although their use may be limited by side-effects, while lower doses of topical steroids are safe and effective for treatment of moderate bullous pemphigoid. IgA-mediated pemphigoid can often be difficult to treat even with usually effective medications such as rituximab.[8-10]

CONCLUSION

Bullous pemphigoid (bull-us pem-fuh-goyd) is a rare skin condition that causes itchy, hive-like welts or fluid-filled blisters.It may affect a small area of your body or it may be widespread. Blisters may occur anywhere, but often develop on flexural areas of your skin, such as under your armpits (axilla), around your groin or on your stomach (abdomen). In some cases, blisters also form on mucous membranes, including your mouth, tongue, throat, esophagus and/or eyes.Bullous pemphigoid appears as itchy welts that look like hives or multiple, itchy blisters (bullae). It most commonly appears on your: Arms, Legs, Abdomen, Groin, Mouth. The blisters typically appear along the creases of your skin. They usually aren’t painful, but they may break open and become a painful sore or ulcer.The fluid inside of your blisters may be clear, or it may contain some blood. The skin around your blisters may look normal or discolored (red, purple, brown or slightly darker than your normal skin color).For mild cases of bullous pemphigoid, the best treatments are topical corticosteroid creams or ointments that you rub directly on the affected areas.For moderate-to-severe cases of bullous pemphigoid, your healthcare provider may prescribe an oral corticosteroid, like prednisone, in addition to a steroid-sparing immunomodulatory agent such as dapsone, mycophenolate mofetil, azathioprine, methotrexate or chlorambucil.If you can’t take can’t corticosteroids or other immunomodulatory agents, your healthcare provider may prescribe

oral tetracycline or doxycycline pills.For refractory cases of bullous pemphigoid, your healthcare provider may prescribe rituximab or IVIG infusions.There aren't any home remedies for bullous pemphigoid, but there are things you can do to make your symptoms more tolerable, including:Wear soft, loose-fitting clothing made out of natural fibers, Avoid spending a lot of time in the sun and wear sunscreen, Wash your sores or ulcers with antibacterial soap and water to prevent infection. Then, apply antibiotic ointment to your affected areas and wrap them in nonadhesive (doesn't stick to your skin) bandages, Avoid standing or walking for long periods if you have bullous pemphigoid on your feet, Moisturize your skin with lotions, creams, coconut oil or shea butter oil.If you have bullous pemphigoid in your mouth, the following tips can make your symptoms more tolerable and help prevent nutrition problems:Eat a diet of soft foods, such as soups, mashed foods, pudding and applesauce.Wait until hot foods cool down before you eat.Avoid crunchy or sharp foods, including chips, cereal, crusty bread and raw vegetables and fruits.Avoid acidic foods, including hot peppers, salsa, citrus fruits and tomatoes.Avoid drinking alcohol.

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