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Bullous Pemphigoid

Bullous pemphigoid (BP) is a chronic blistering of the skin. It ranges from mildly itchy welts to severe blisters and infection, and may affect a small area of the body or be widespread. The vast majority of those affected are elderly, but it has been seen at all ages.

It is an autoimmune disorder, meaning it is caused when the body's immune system malfunctions. The immune system is meant to defend the body against bacteria, viruses, and disease, but instead produces antibodies against healthy tissue, cells and organs. Some patients with BP have other autoimmune diseases such as diabetes and rheumatoid arthritis. Various other factors have been reported to play a role in triggering BP. These include drugs (furosemide, penicillins) and mechanical trauma and physical traumas (burns from radiation, sun or heat).

Bullous is the medical term for a large blister (a thin-walled sac filled with clear fluid). Usually the skin in BP is very itchy and large, red welts and hives may appear before or during the formation of blisters. The blisters are widespread and usually appear on the areas of the body that flex or move (flexural areas).

Variations of BP include localized pemphigoid (covers a small area but heals with scars), Pemphigoid vegetans (large oozing area in the groin and armpits), cicatricial pemphigoid (scarring form of the mouth and eyes), vesicular pemphigoid (eczema-like rash), urticarial pemphigoid (hive-like), erythrodermic pemphigoid (entire skin redness) and pemphigoid nodularis (insect bite-like sores that don't heal). About 15-20 percent of people with BP also develop blisters in the mouth, throat, or esophagus.

Because of all the variations and differing degrees of symptoms, the diagnosis must be confirmed by skin biopsy. A special skin biopsy test (a direct immunofluorescence biopsy) may also be needed. Blood tests are usually inconclusive.

Treatment is focused on relief of symptoms and prevention of infection. Tetracycline and Minocycline antibiotics are very useful for mild to moderate disease. They do not work on bacteria, but act directly on the immune system. They can be used in combination with potent topical steroid creams for more rapid relief.

Oral steroids (prednisone, prednisolone) are the treatment of choice for severe cases. Regular visits will be needed because the dose must be adjusted frequently, and side effects must be monitored. A fairly high dosage is needed initially, and once the blisters have stopped appearing, it is slowly reduced over many months or years. As steroids have some undesirable side effects, we try to reduce the dose as much as possible. If this is done too quickly, the blisters reappear.

Often, immunosuppressive agents (Immunan, Cellcept, Methotrexate, cyclophosphamide and Neoral) are used in combination with the oral steroids to allow a lower dose. Severe cases are best treated in the hospital to allow expert dressing of the wounds, and intravenous injections of the most potent treatments.

BP is a self-limiting disease that in most cases eventually completely clears up, so that the treatment can be stopped. Treatment is usually needed for several years, but generally after a few months it is possible to reduce the dose of medications to reasonably low levels. BP also often has a pattern of remissions and flare-ups. It may be without symptoms for 5 or 6 years then suddenly flare up.

With careful management, most patients with BP do well. Be patient and faithfully follow your instructions. These are the keys to good results.