PEMPHIGUS

http://www.aocd.org

Pemphigus is a rare autoimmune blistering disorder of the skin. The immune system produces protective antibodies that circulate through our blood and protect us from attack by hostile viruses and bacteria. In pemphigus however, the antibodies mistakenly consider normal tissues as foreign and attack them. This produces painful raw areas on the skin and mucous membranes (mouth and other moist linings) that will not heal. In some cases, these sores can cover a significant area of the skin. Before modern drug treatments, death from overwhelming infection was the usual outcome, but this is no longer common.

Pemphigus has several types. Each type looks and acts differently. There are other autoimmune blistering diseases of the skin, such as **bullous pemphigoid**, bullous lupus, and **Hailey-Hailey disease** that can be confused with pemphigus. Because this is a complex condition to diagnose, special testing is needed. The three main categories of pemphigus are paraneoplastic pemphigus, pemphigus vulgaris and pemphigus foliaceus.

This most serious form of pemphigus is paraneoplastic pemphigus. This occurs most often in someone who has already been diagnosed with an internal malignancy (cancer). Fortunately, it is also the most rare. Painful sores of the mouth, lips and esophagus are almost always present; and skin lesions of different types occur. This disease is usually not responsive to treatment. The diagnosis of this type should prompt doctors to search for a hidden tumor. In some cases the tumor will be benign and the disease will improve if the tumor is surgically removed.

Pemphigus vulgaris is the most frequently diagnosed form of pemphigus. Sores and blisters usually start in the mouth. The sores don't always look like blisters. Pemphigus vulgaris is produce by antibodies in the blood that attack the skin directly. The antibodies circulate in the blood, reach the skin and bind to a particular protein found on skin cells. This protein's normal function is to keep the skin cells bound together. When the antibodies bind the protein, the cells fall apart and blistering of the skin and mucous membranes occur.

With pemphigus foliaceus crusted, scaly sores, or fragile blisters usually first appear on the scalp and then later involve the face, chest and back. The antibodies bind to a different skin cell protein than in pemphigus vulgaris. This protein is found only on the top dry layer of skin, and is not found in the mouth. Because of this the blisters are superficial, and do not occur in the mouth. They are often itchy and not usually as painful as pemphigus vulgaris.

Pemphigus vulgaris usually starts in middle-aged and older adults, but is can occur at any age. Often, pemphigus not the first disease considered. The first line of defense for all forms of pemphigus is the introduction of **oral steroids**, usually Prednisone. **Cytotoxic drugs** such as Imuran and Cytoxan are added to reduce the dose and side effects of the steroids. Other drugs used are injectable gold and **cyclosporin**. Mild disease can sometimes be controlled with **topical** or **intralesional steroids**. **Hydroxychloroquine** (Plaquenil) and **dapsone** have also have been effective in some cases of pemphigus foliaceus.

All of these drugs can have serious side effects. Blood and urinalysis must be monitored on a regular basis. There is some evidence that treatment is easier in the early stages of the disease. Treatment should always be prescribed on the basis of how severe the disease is. There are no reports showing that alternative, homeopathic or herbal treatments help. These usually hurt because the medications that work are stopped until the patient realizes they made a mistake. This same pattern is seen over and over, usually in intelligent and more educated patients.

This information has been provided to you compliments of the American Osteopathic College of Dermatology and your physician.