Close-up on: Pemphigus

By Shweta Dhawan and Elena Netchiporouk

Pemphigus is the name for a group of rare but potentially life-threatening blistering autoimmune diseases that affect the skin and mucous membranes, such as those found in the mouth. It is caused by antibodies (immune proteins) in the body mistakenly attacking the cement that holds skin cells together. As a result, the skin cells are no longer held together and blisters, erosions and small pus-filled bumps can develop.

There are two main types of pemphigus: pemphigus foliaceus and pemphigus vulgaris. Pemphigus foliaceus usually leads to very superficial itchy erosions, and blisters are rarely seen. Pemphigus vulgaris, on the other hand, presents with blisters and erosions in the mouth or other mucous membranes, and sometimes on the skin. This is the most common form of pemphigus in North America and Europe.

Rarer subtypes of pemphigus can arise from the side effects of medication or from diseases of the internal organs (e.g., immunoglobulin A pemphigus and paraneoplastic pemphigus). *See your doctor if you have blisters inside your mouth or on your skin that do not heal.*

How does pemphigus develop and who gets it?

Our bodies make antibodies to fight infections. Pemphigus is an autoimmune disorder in which the body produces antibodies that damage the cells of the skin and mucous membranes. It is a result of an interaction between genetics and external factors, such as an environmental agent (probably an unidentified microbe). Occasionally, medications can induce this condition and it resolves when the drug is discontinued. The disease often affects middle-aged adults. It is not contagious.

How is pemphigus diagnosed?

Several conditions can cause blisters, and a diagnosis can, therefore, be difficult to determine. Your doctor will ask about your symptoms, examine your skin and mouth, and conduct blood tests, and might refer you to a dermatologist.

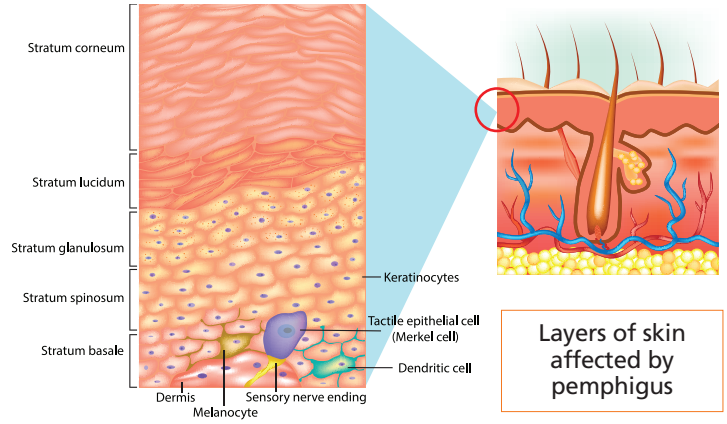
A diagnosis of pemphigus is confirmed by two biopsies—one of the blister and the other of the skin adjacent to it. The tissue from the blister is examined under a microscope to confirm the level of the skin that is affected by the blister. The tissue from the skin adjacent to the blister is sent for specific tests, called immunofluorescence studies, to identify the autoantibodies involved.

How is pemphigus treated?

Pemphigus can be life-threatening because having fragile skin makes you vulnerable to severe infections. Mild pemphigus can be treated with topical medications, such as a mouthwash containing a mixture of corticosteroids, numbing agents and antimicrobials.

Most people need to take oral corticosteroids to help the blisters heal faster and prevent new lesions. Corticosteroids work very fast, but have many adverse effects when taken orally for a long time. For this reason, your doctor may give you a different medication to help decrease the production of antibodies and that is safer to use for several months or years. The most common treatments are oral medicines such as mycophenolate mofetil and drugs that are infused into the blood, such as rituximab or intravenous immunoglobulins. Because these medicines are potentially toxic, your dermatologist may ask you to have regular blood tests. A regular follow-up with your dermatologist and careful oral hygiene are important for the treatment to be successful.

Pemphigus unfortunately tends to be a long-lasting condition, but medications can usually control it. CS



Layers of skin affected by pemphigus

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