CONDITION

Brazilian pemphigus foliaceus

Learn about Brazilian pemphigus foliaceus, an autoimmune blistering skin disease, including its symptoms, causes, diagnosis, treatment, and follow-up care.

WHAT IT IS
Rare autoimmune blistering skin disease

BODY SYSTEM
Skin

MAIN SYMPTOMS
Fragile blisters, scaly or crusted patches, raw areas, burning pain

WHAT CAUSES IT
Autoimmune, with inherited and environmental susceptibility

HOW IT IS DIAGNOSED
Skin biopsy and direct immunofluorescence

MAIN TREATMENT
Corticosteroids and immune-suppressing medicines

CAN IT BE CURED?
No, relapses can occur after improvement

IS IT AN EMERGENCY?
Sometimes, rapidly spreading skin peeling with fever

LONG-TERM OUTLOOK
Often controllable, with relapses and ongoing follow-up

Overview

Brazilian pemphigus foliaceus is a rare autoimmune blistering disease that affects the outer layers of the skin. It is also called fogo selvagem , a Portuguese term meaning "wild fire." The name refers to the burning feeling that some people experience and to the disease's occurrence in certain rural regions of Brazil.

In an autoimmune disease, the immune system mistakenly attacks part of the body. In Brazilian pemphigus foliaceus, the immune system produces antibodies against desmoglein 1 , a protein that helps skin cells stick together. When this connection is disrupted, cells near the skin's surface separate. This causes fragile blisters, peeling, crusting, and raw areas called erosions.

The blisters are so delicate that they often break before they can be seen. A person may notice scaly or crusted patches rather than intact, fluid-filled blisters.

Unlike pemphigus vulgaris, another form of pemphigus, Brazilian pemphigus foliaceus usually does not cause sores inside the mouth or other moist body surfaces. This is related to how desmoglein proteins are distributed in the skin and mucous membranes.

The condition is not contagious. It cannot be spread through touch, shared belongings, food, or air. It is also not caused by poor hygiene.

Brazilian pemphigus foliaceus matters because untreated disease can become widespread. Extensive skin damage may lead to pain, fluid loss, difficulty controlling body temperature, or infection. Treatment can often control the immune attack and allow the skin to heal, but ongoing follow-up is usually needed.

Symptoms

Symptoms may begin in one area and gradually spread. The scalp, face, chest, and upper back are common early sites because these areas contain many oil-producing glands. Some people develop more generalized skin involvement.

Possible symptoms include:

The skin may resemble eczema, seborrheic dermatitis, psoriasis, or a bacterial skin infection. Because intact blisters may be absent, the condition can be difficult to recognize from appearance alone.

Mouth sores are unusual in pemphigus foliaceus. New sores involving the mouth, eyes, nose, genitals, or throat should be reported to a clinician because they may point to another condition or a different form of pemphigus.

Symptoms may improve for a time and later return. This is called a relapse. The pattern varies from person to person and may be affected by treatment, sun exposure, infections, or other immune triggers.

- Fragile, superficial blisters that quickly break
- Red, scaly, or crusted patches
- Raw or weeping areas where the top layer of skin has peeled away
- Burning, tenderness, or pain
- Itching
- Flaking or thick crusts on the scalp, face, chest, or back
- Skin that peels or forms an erosion after rubbing or pressure

- Worsening after sun exposure or heat
- Widespread redness and scaling in more severe disease

Causes and risk factors

Brazilian pemphigus foliaceus develops when the immune system makes immunoglobulin G, or IgG, antibodies that target desmoglein 1. This protein acts like part of the "glue" holding skin cells together. Damage to it causes the cells to separate, a process called acantholysis .

Why this immune reaction starts is not fully understood. Current evidence suggests that inherited susceptibility and environmental exposure may work together.

The disease has been found in clusters in certain rural and Indigenous communities in Brazil, especially in areas near rivers and streams. Research has identified an association with exposure to bites from particular black flies, including Simulium nigrimanum . Scientists have proposed that proteins in insect saliva may trigger or strengthen an immune response in people who are genetically susceptible. This connection remains an area of study; an insect bite by itself does not mean that someone will develop the disease.

Risk factors and associations include:

Most people exposed to black flies do not develop pemphigus. The condition is not passed directly from parent to child, although inherited traits may affect susceptibility.

Some medicines have been linked to pemphigus foliaceus outside endemic settings. A clinician will review prescription drugs, over-the-counter products, and supplements when evaluating symptoms. People should not stop a prescribed medicine unless the prescriber advises them to do so.

- Living in or spending substantial time in an endemic rural area
- Frequent exposure to biting insects in affected regions
- Certain inherited immune-system traits, including specific HLA gene patterns
- Having relatives or community members with the disease, reflecting shared genes and environment
- Younger age than is typical for non-endemic pemphigus foliaceus

Diagnosis

Diagnosis usually involves a dermatologist and combines the skin examination with laboratory

testing. Appearance alone is not enough because several other skin conditions can look similar.

A clinician will ask when the rash began, where it first appeared, and whether it burns, itches, or worsens in sunlight. They may ask about residence or travel in rural Brazil, insect exposure, family history, recent infections, and medicines.

A skin biopsy is central to diagnosis. After numbing the area, the clinician removes a small piece of skin for examination under a microscope. In pemphigus foliaceus, the pathologist may see separation of skin cells just below the outermost layer.

A second biopsy may be taken from normal-looking skin next to a fresh lesion. It is tested by direct immunofluorescence , a method that uses fluorescent markers to show immune deposits in tissue. Pemphigus typically produces a net-like pattern of IgG, sometimes with a complement protein called C3, around skin cells.

Blood tests may look for antibodies against desmoglein 1. An enzyme-linked immunosorbent assay, or ELISA, can help support the diagnosis and may sometimes help track disease activity. Antibody levels must be interpreted alongside symptoms and biopsy findings. Some people living in endemic areas may have low-level antibodies without active skin disease.

Other conditions that may need to be ruled out include bacterial infections such as bullous impetigo, seborrheic dermatitis, lupus-related skin disease, other autoimmune blistering disorders, medication reactions, and staphylococcal scalded skin syndrome.

Treatment and management

Treatment aims to stop new blisters, heal existing areas, reduce discomfort, and prevent complications. The plan depends on how much skin is affected, how quickly the disease is spreading, other health conditions, and previous treatment responses.

Limited disease may sometimes be managed with corticosteroid medicine applied to the skin. These medicines reduce inflammation and immune activity. Large or sensitive areas require careful supervision because strong topical corticosteroids can be absorbed through damaged skin and may cause side effects.

More extensive disease often requires treatment that works throughout the body. Oral corticosteroids can control the disease relatively quickly, but long-term use may cause infection risk, high blood sugar, high blood pressure, bone loss, cataracts, mood changes, and other problems. Clinicians generally try to use the lowest effective exposure and reduce treatment gradually when disease control allows. Corticosteroids like prednisone or hydrocortisone should not be stopped suddenly without medical guidance.

Other immune-suppressing medicines may be used to reduce reliance on corticosteroids. Options described in pemphigus guidance include azathioprine and mycophenolate mofetil. Rituximab, an antibody treatment that targets immune cells called B cells, may be considered for significant or difficult-to-control pemphigus. Dapsone and other therapies may be appropriate in selected cases. The choice requires individualized assessment, screening, and laboratory monitoring.

Before immune-suppressing treatment, clinicians may check for infections such as hepatitis or tuberculosis, depending on the planned medicine and the person's risk factors. Vaccination needs may also be reviewed. Live vaccines may not be appropriate during some treatments, so timing should be discussed with the care team.

Supportive skin care is also important:

Antibiotics like amoxicillin or doxycycline are not routine treatment for pemphigus itself. They may be prescribed if a bacterial infection is confirmed or strongly suspected.

- Clean affected areas gently and follow wound-care instructions.
- Use non-stick dressings when advised.
- Avoid rubbing, harsh soaps, and adhesive products on fragile skin.
- Protect the skin from excessive sunlight with shade, clothing, and suitable sunscreen.
- Ask how to manage pain, itching, or scalp crusting safely.
- Use insect protection in endemic areas, while avoiding products that irritate damaged skin.
- Maintain adequate food and fluid intake if large areas are affected.

Complications

Loss of the skin barrier can make it easier for bacteria to enter the body. Warning signs of a local infection include increasing warmth, swelling, tenderness, pus, unpleasant odor, or redness spreading beyond an affected area. Fever or worsening weakness may indicate a more significant infection.

Widespread skin loss can also lead to fluid loss, changes in body temperature, and problems maintaining nutrition. Pain, visible skin changes, and long treatment courses may affect sleep, work, relationships, and emotional health.

Treatment can cause complications as well. Immune-suppressing medicines increase susceptibility to certain infections. The specific risks vary by medicine. Regular blood tests

may be needed to monitor blood-cell counts, liver function, kidney function, or other measures. Long-term corticosteroid treatment may require attention to bone health, blood pressure, blood sugar, and eye health.

Seek urgent medical advice for rapidly worsening skin disease, new fever, spreading redness, pus, reduced urination, dizziness, or new sores involving the eyes, mouth, throat, or genitals.

Emergency care is appropriate for trouble breathing, facial or throat swelling, fainting, confusion, severe weakness, or extensive and rapidly spreading skin peeling accompanied by fever or severe pain.

Follow up

Regular dermatology follow-up helps determine whether treatment is preventing new lesions and whether healing is continuing. Visits may be frequent when treatment starts or changes. Once the disease is stable, appointments may be spaced farther apart.

The clinician may review the number of new blisters, the area of involved skin, discomfort, infection signs, and possible treatment side effects. Photographs or standardized disease scores may be used to track changes. Blood antibody levels can sometimes add useful information, but treatment decisions are not based on a laboratory result alone.

Laboratory monitoring depends on the medicine being used. It may include complete blood counts and tests of liver or kidney function. People taking corticosteroids for an extended period may also need monitoring for blood pressure, blood sugar, bone loss, and eye problems.

Report a return of blisters or crusted areas rather than changing treatment independently. A relapse does not necessarily mean that all treatment has failed. The care team may reassess triggers, confirm that infection is not present, and adjust the management plan.

Routine appointments are also a good time to discuss sun protection, insect-bite prevention, vaccination timing, pregnancy plans, medication interactions, and the emotional effects of living with a visible skin condition.

Sources

- Hans-Filho G, Aoki V, Bittner NRH, Bittner GC. "Fogo selvagem: endemic pemphigus foliaceus." Anais Brasileiros de Dermatologia . 2018;93(5):638-650.



- Murrell DF, Peña S, Joly P, et al. "Diagnosis and management of pemphigus: Recommendations of an international panel of experts." Journal of the American Academy of Dermatology . 2020;82(3):575-585.e1.
- Joly P, Horvath B, Patsatsi A, et al. "Updated S2K guidelines on the management of pemphigus vulgaris and foliaceus initiated by the European Academy of Dermatology and Venereology." Journal of the European Academy of Dermatology and Venereology . 2020;34(9):1900-1913.
- Culton DA, Qian Y, Li N, et al. "Advances in pemphigus and its endemic pemphigus foliaceus phenotype, fogo selvagem: A paradigm of human autoimmunity." Journal of Autoimmunity . 2008;31(4):311-324.
- Stanley JR, Amagai M. "Pemphigus, bullous impetigo, and the staphylococcal scalded-skin syndrome." New England Journal of Medicine . 2006;355(17):1800-1810.