EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

Information Leaflet for Patients

Mucous Membrane Pemphigoid

The aim of this leaflet

This leaflet is designed to help you understand more about mucous membrane pemphigoid. It tells you what this condition is, what causes it, how it is diagnosed and treated, and practical advice for managing this condition.

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Mucous Membrane Pemphigoid

What is mucous membrane pemphigoid?

Mucous membrane pemphigoid is a chronic autoimmune disease. It typically occurs in the elderly, with a mean age between 60 and 80 years old. With about 1 to 2 newly diagnosed patients per million individuals per year in Western Europe, mucous membrane pemphigoid is a rare disease.

What does mucous membrane pemphigoid look like, and what are the signs and symptoms?

As the name suggests, lesions in mucous membrane pemphigoid occur predominantly or exclusively on one or more mucous membranes. Mucous membrane pemphigoid most frequently affects the mouth, the conjunctiva of the eyes and the nasal mucosa, followed by the genital region, the throat and the. The signs and symptoms depend on the affected mucous membrane. When the mouth is affected, red patches, blisters or *erosions* (mucosa breakdown) of the gums, mucosa of cheeks, palate and tongue can be seen. The most common symptoms of the mouth include discomfort, burning, bleeding of the gums, mucosal peeling and difficulty eating. The signs of affected eyes include redness, tearing, burning, decreased vision and foreign body sensation. Coughing and voice modification can occur when the throat (larynx) is affected, and painful swallowing when the oesophagus is affected. Patients with genital involvement suffer from pain, itch and burning accompanied by redness, blisters and *erosions* (mucosa breakdown). The skin can be mildly affected with blisters, erosions and crusts (scabs) when damaged. The lesions may progress by forming scars that sometimes permanently damage the affected mucosa. These scars sometimes have serious consequences, especially in the eyes and throat (larynx). Scarring of the eyes can cause blindness and scarring of the throat can cause shortness of breath.

What is the cause of mucous membrane pemphigoid?

Our immune system produces specialized proteins called *antibodies*, which bind to bacteria, viruses, fungi (the plural of fungus), and tumor cells, and protect us from these infections as well as cancer. In patients with mucous membrane pemphigoid, the immune system mistakenly produces *antibodies* that bind to certain structures in the mucous membranes and skin. These structures are termed BP180, BP230, laminin 332, and type VII collagen and provide attachment between the upper layers of the skin and mucous membranes from the deeper layers. These disease-causing *antibodies* that bind to the mucous membranes and skin are called *auto-antibodies* (*antibodies* against one's own body). This binding triggers a complex inflammatory reaction that finally leads to the separation of the upper layer of the mucous membranes and skin from the deeper layers. So far, it is unclear what factor(s) trigger(s) the immune system to produce the disease-causing *auto-antibodies*.

How is mucous membrane pemphigoid diagnosed?

The clinical characteristics give important clues to the diagnosis of mucous membrane pemphigoid. However, for a specific diagnosis, the detection of auto-antibodies is required. These auto-antibodies can be detected in the mucous membranes and the skin through a mucosal and/or skin biopsy (usually 4 mm in diameter). This procedure is done

under local anesthesia in a few minutes. For the detection of auto-antibodies in the blood, a blood sample is taken.

How does mucous membrane pemphigoid develop?

Mucous membrane pemphigoid usually progresses to become a chronic disease, and treatment may be required for months and sometimes years. During this time, new blisters and erosions can appear on one or more mucous membranes. For this reason, regular follow-up visits, ideally in a specialized outpatient department, are recommended to allow dermatologists to assess disease activity, adapt the medication, check for possible side effects, and measure the blood levels of auto-antibodies. In particular, affection of the eyes can lead to scarring of the eye tissue followed by vision impairment and ultimately, blindness.

What are risk factors and associated diseases?

Some medications, such as gliptins used in the treatment of diabetes and checkpoint inhibitor therapy, a form of cancer immunotherapy, may trigger the onset of mucous membrane pemphigoid. If mucous membrane pemphigoid is diagnosed, these drugs may be replaced, or in case of the cancer immunotherapy, continued if possible. In 10-20% of patients with mucous membrane pemphigoid, auto-antibodies react with a specific protein in the skin/ mucous membranes, termed laminin 332. In about a quarter of patients with auto-antibodies reactive with laminin 332, a cancer can be found. In this patient subgroup, a tumor search is recommended.

How is mucous membrane pemphigoid treated?

Since mucous membrane pemphigoid can affect several mucous membranes, the treatment will involve several specialists. The dermatologist generally coordinates the care and depending on the affected mucosal sites, other specialists such as the ophthalmologists, oto-rhinolaryngologists and gynaecologists can be involved. In general, therapeutic options include local (e.g., mouthwashes, eye drops or ointments and creams) and systemic (oral) treatments (tablets), often in combination. In moderate to severe disease, anti-inflammatory drugs (like dapsone) or immunosuppressants (drugs that block the production of autoantibodies) such as methotrexate, cyclophosphamide, mycofenolate mofetil, azathioprine, and rituximab are given. Cortisone tablets may be necessary until the above-mentioned treatments become effective.

Can mucous membrane pemphigoid be prevented?

So far, no preventive measures to avoid the occurrence of mucous membrane pemphigoid are known.

What is practical advice for taking care of mucous membrane pemphigoid?

- Mucous membrane pemphigoid is a rare disease and is best treated in a specialized clinic experienced with this disorder.
- Since some drugs may interfere with those prescribed for mucous membrane pemphigoid, please inform your dermatologist about any new drug prescribed since your last visit.
- In case of involvement of the mouth, careful oral hygiene and appropriate care are recommended.

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Patient support groups

There are country-specific and international patient support groups for patients suffering from mucous membrane pemphigoid, and may help you or your friend/family member find further help.

connection between arteries and veins) in their early phase as another possible diagnosis.

Patient support organisations

UK: www.pemfriends.co.uk and www.pemphigus.org.uk

Germany: www.pemphigus-pemphigoid-selbsthilfe.de

Italy: www.assoc-apai.org and www.pemfigo.it

France: www.pemphigus.asso.fr

Spain: viviconpenfigo.blogspot.com

Turkey: www.turkdermatoloji.org.tr

International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

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