EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

Information Leaflet for Patients

Paraneoplastic pemphigus

The aim of this leaflet

This leaflet is designed to help you understand more about paraneoplastic pemphigus. 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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INFORMATION LEAFLET FOR PATIENTS 2022

Paraneoplastic pemphigus

What is paraneoplastic pemphigus?

Paraneoplastic pemphigus is an autoimmune disease that mainly affects the mouth and skin. Other mucosal tissues such as throat, eyes, and genital areas can also been involved and in some patients, also the airways and lungs. Paraneoplastic pemphigus is typically associated with hematological malignancies (blood cancer), more rarely, with solid tumors. Paraneoplastic pemphigus is a very rare disease and there is limited data on its frequency.

What does paraneoplastic pemphigus look like, and what are the signs and symptoms?

The spectrum of mucocutaneous lesions in paraneoplastic pemphigus is broad. Typically, severe lesions in the mouth, on the tongue and on the lips occur (**Fig. 1**). Skin lesions can be variable and manifest as blisters and erosions that may mimic other autoimmune bullous diseases, as erythema (reddish skin) or violaceous papules and plaques resembling lichen planus (**Fig. 2**). Paraneoplastic pemphigus may also mimic severe diseases like toxic epidermal necrolysis or Stevens Johnsons syndrome. The respiratory tract can be affected with bronchiolitis obliterans representing one of the leading causes of death in these patients.

During the course of the disease lack of appetite, weight loss, fever, skin and systemic infections, and general weakness can develop.

What causes the blistering?

Our immune system produces specialised proteins called antibodies, which bind to bacteria,

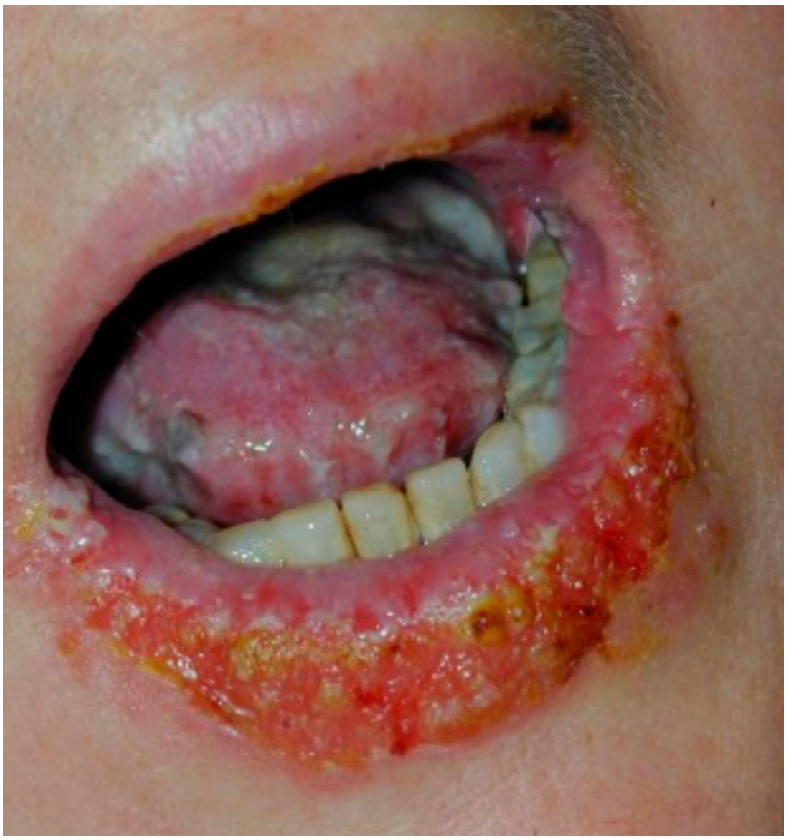

Fig. 1 *Erosive oral mucositis*

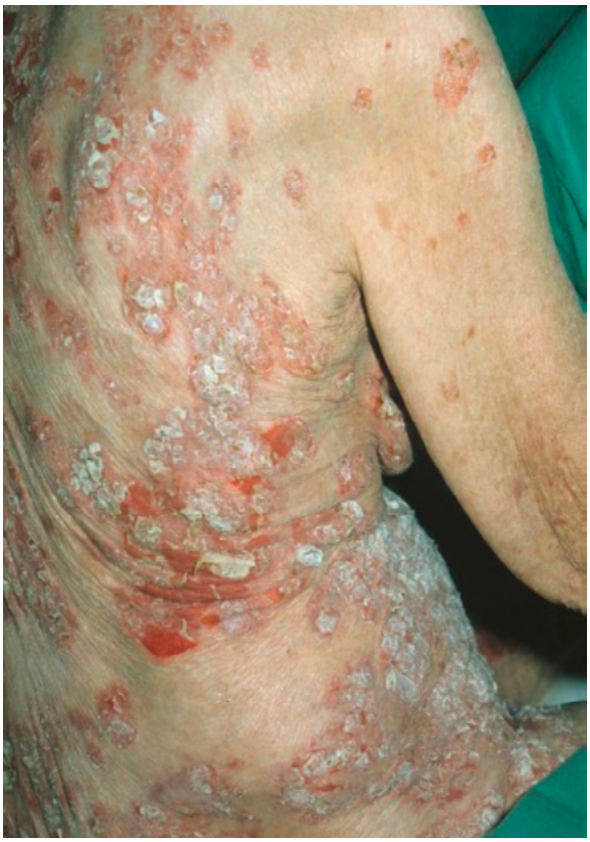

Fig. 2 *Erythematous* (reddish) squamous lesions associated with erosions on the back

viruses, fungi (the plural of fungus) and tumor cells, thus protecting us both from infections and cancer. In patients with paraneoplastic pemphigus, the immune system mistakenly produces antibodies that bind to certain structures in the skin and mucous membranes. These disease-causing antibodies are called auto-antibodies (antibodies against one's own body). This binding triggers a complex inflammatory reaction that finally leads to the cell detachment and erosion sin skin and mucous membranes.

How is paraneoplastic pemphigus diagnosed?

Clinical characteristics alone can give important clues for the diagnosis of paraneoplastic pemphigus. However, additional diagnostic steps are required. The detection of auto-antibodies against different autoantigens is essential. These auto-antibodies can either be detected in the skin (**Fig. 3**) or in the blood. To detect auto-antibodies in the skin, a skin biopsy (usually 4 mm in diameter) is performed. This procedure is done under local anesthesia and only takes a few minutes. For the detection of auto-antibodies in blood, a blood sample is taken. Finally, a comprehensive screening workup is mandatory in order to disclose a possible underlying malignancy.

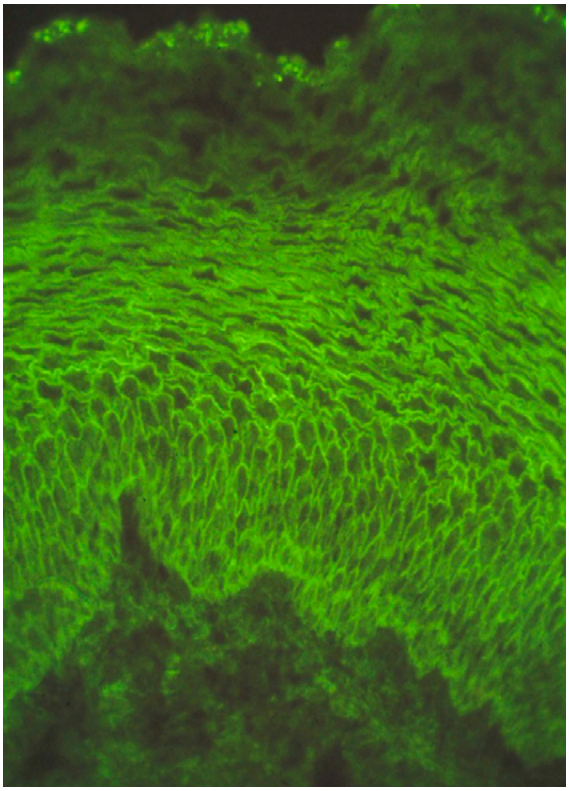

Fig. 3 immunoglobulin G (IgG) *auto-antinodies* can be detected in netlike or 'chicken wire' pattern in a skin biopsy (by direct uimmunofluorescence microscopy)

How does paraneoplastic pemphigus develop?

Paraneoplastic pemphigus is a severe condition with a high mortality rate. During the natural disease course of PNP, new blisters and erosions can appear.

What are the risk factors and associated diseases?

Some malignancies are associated with paraneoplastic pemphigus more frequently than others. In particular, hematological malignancies (mainly non-Hodgkin lymphoma, chro¬nic lymphocytic leukemia, Castleman's disease and thymomas) account for the vast majority of paraneoplastic pemphigus cases. Your dermatologist or general physician should rule out those malignancies.

How is paraneoplastic pemphigus treated?

In general, therapeutic options include systemic (oral) treatments in the form of tablets, which may be combined with local therapies (e.g., ointments and creams). The treatment of the underlying malignancy is recommended. Usually the systemic treatment consists of cortisone tablets combined with immunosuppressants (such as methotrexate, azathioprine or mycophenolate) may also be administered. Also, targeted therapies such as anti-CD20 monoclonal antibody (rituximab) have been successfully used in some patients with refractory disease.

Paraneoplastic pemphigus

Can paraneoplastic pemphigus be prevented?

So far, no preventive measures to avoid the occurrence of paraneoplastic pemphigus are known.

What is practical advice for taking care of paraneoplastic pemphigus?

- Paraneoplastic pemphigus is a very rare disease and needs to be treated in a clinic specialized for this condition.
- Since some drugs may interfere with those prescribed for paraneoplastic pemphigus, please inform your dermatologist about any new drug prescribed since your last visit.
- There are country-specific and international support groups for patients suffering from paraneoplastic pemphigus, which may help you or your friend/family member receive further assistance.

Patient support organisations

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

Germany: www.pemphigus-pemphigoid-selbsthilfe.de

Italy: www.pemfigo.it

France: www.pemphigus.asso.fr

Turkey: www.turkdermatoloji.org.tr

International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

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