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Paraneoplastic pemphigus

What is paraneoplastic pemphigus?

The pemphigus family are rare autoimmune blistering diseases affecting skin and/or mucous membranes. Paraneoplastic conditions occur in association with malignancies (cancer). Paraneoplastic pemphigus is characterised by painful blisters and denuded areas of the mouth, lips, oesophagus and skin. It is the least common but most serious form of pemphigus, particularly considering the presence of the underlying cancer.

What causes paraneoplastic pemphigus?

An autoimmune disorder is a disease in which an individual's immune system starts reacting against his or her own tissue.

The building block cells of the epidermis are called keratinocytes. These cells are cemented together at special sticky spots called desmosomes. In paraneoplastic pemphigus and other types of pemphigus there are autoantibodies to a component of the desmosomes. These cause the keratinocytes to separate from each other. The gap fills with fluid, causing a blister or the skin may peel off leaving raw areas.

What are the signs and symptoms of paraneoplastic pemphigus?

Almost all patients with paraneoplastic pemphigus have an associated malignancy. [Oral blisters](#), erosions and ulcerations may be very painful. Other mucosal surfaces affected include the nose, throat and genitals. Skin lesions occur anywhere on the body and are highly variable in appearance. They can be red and inflamed spots, scaly plaques, fluid-filled blisters or ulcerative lesions.

It may be confused with several other blistering skin conditions including [pemphigus vulgaris](#), [erythema multiforme](#), [bullous pemphigoid](#), and [lichen planus](#).

Paraneoplastic pemphigus may also affect the respiratory and gastrointestinal tracts, which significantly increases the chances of death.

Tests for paraneoplastic pemphigus

Diagnosis of paraneoplastic pemphigus requires a [skin biopsy](#), which shows rounded-up separated keratinocytes (called acantholytic cells) within the blisters, dead keratinocyte cells and an inflammatory reaction. Direct immunofluorescence staining of the skin sections reveals antibodies targeted at desmoplakins. These are proteins in the desmosome complex.

In some cases, circulating immunoglobulin type G (IgG) antibodies can be detected by a blood test (indirect immunofluorescence test).

Treatment of paraneoplastic pemphigus

Confirmed diagnosis of paraneoplastic pemphigus will prompt the doctor to look for a hidden tumour if the patient is not already known to have one. The condition can resolve if the cancer can be removed or cured. Treatment is otherwise difficult and largely just supportive, aimed at dressing the areas, pain relief and managing secondary infections. 75–80% of patients die from paraneoplastic pemphigus or from the underlying cancer.

Related information

References:

On DermNet NZ:

- [Blistering diseases](#)
- [Pemphigus vulgaris](#)
- [Pemphigus foliaceus](#)
- [Bullous pemphigoid](#)
- [Oral blistering diseases](#)

Other websites:

- [Pemphigus, paraneoplastic](#) - emedicine dermatology, the online textbook

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DermNet does not provide an on-line consultation service.
If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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