

Pemphigus

Pemphigus is a life-threatening recurrent autoimmune disease with intraepidermal blisters production on the skin and on the mucous membranes, which mostly affects older people, between 30 and 60 years.

Pemphigus Vulgaris

An acquired, chronic autoimmune disease. In primary blisters formation on mucous membranes, it is possible to detect IgG against keratinocyte membrane antigens, desmoglein 3, in primary skin form - against desmoglein 1. Direct immunofluorescence allows the detecting of IgG and C3 bound in IC slots of the epidermis.

Clinical picture

Intraepidermal **vesicle or bulla**, without any significant predilection – later it comes to generalisation. These changes are determined by acantholysis. The blisters have a tight cover and pure content. Later it becomes turbid, the area around is inflamed, a blister burst and leads to the erosion. The erosion gets wet, dirty brown greasy crusts appear on its surface and then begin to smell bad. The skin around is also eroded - it looks normal, but it's possible to wipe it away with mild pressure up to the erosion – Nikolsky's sign, acantholysis also determines indirect Nikolsky's sign. Pruritus does not occur in the skin, erosion burns and hurts, epithelialization is slow and in the end, it heals without scar formation. Later the new blisters are formed coming **in attacks**. **More malignant** forms can cause death. Nowadays patients die rather from the consequences of treatment (cachexia or intercurrent illness) than from the disease itself. Disease on the mucous membranes has a similar course and clinical picture – salivation, foetor ex ore. The illness occurs mostly in patients aged from 40 to 70 years^[1].

Differential diagnosis

Mainly bullous pemphigoid and dermatitis herpetiformis Duhring.

Histology

Intraepidermal blister with **acantholysis** and acanthotic cells (pyknosis).

Prognosis

All forms are severe, with unfavourable prognosis, patients usually die after several years – cachexia, infection.

Treatment

Treatment is only symptomatic - corticosteroids, ACTH, immunosuppressive drugs, ATB.

Pemphigus vegetans

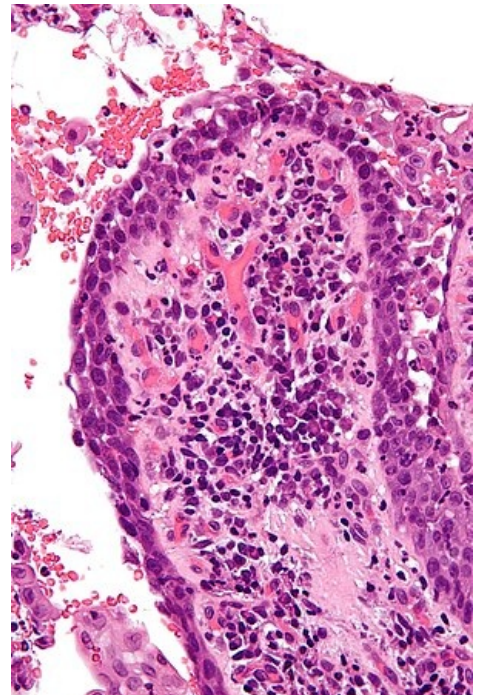
Is milder and more chronic, can transform into vulgaris and regress, is intertriginous, bottoms of the blisters spread in red vegetations covered by the smelly detritus, merge in the verrucous areas, often secondarily infected by the yeasts.

Pemphigus foliaceus

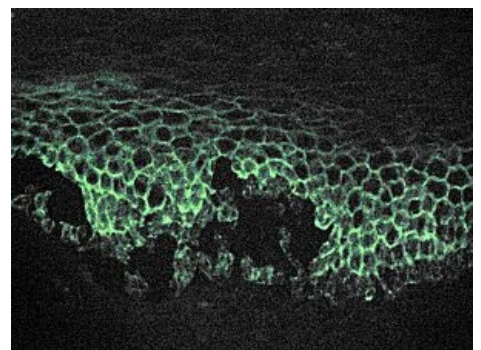
Affects the whole body skin in middle-aged people in form of the exfoliative erythroderma. Acantholysis is subcorneal.^[1]

Wet and smelly crustasquamas are formed in the seborrheic predilection, the mucosa is usually not affected, and itchy.

Pemphigus erythematosus



Pemphigus Vulgaris – loss of cohesiveness of keratinocytes (**acantholysis**) with cracks and blisters formation



Immunofluorescence of the pemphigus shows the accumulation of the antibodies around the desmosome

It's a bounded and milder form of the pemphigus which is characterized by the formation of symmetric, nummular deposits in the seborrheic localization covered by firmly adhering crustasquamas.

Pemphigus herpetiformis

Reminds rather a dermatitis herpetiformis or pemphigoid, products strongly itching blisters.

Links

Related articles

- Autoimmunity
- Skin
- Pemphigoid

Source

- BENEŠ, Jiří. *Studijní materiály* [online]. [cit. 2010]. <<http://jirben.wz.cz>>.
- POVÝŠIL, Ctibor – ŠTEINER, Ivo – DUŠEK, Pavel, et al. *Speciální patologie*. 2. edition. Praha : Galén, 2007. 430 pp. pp. 394. ISBN 978-807262-494-2.
- ŠTORK, Jiří, et al. *Dermatovenerologie*. 1. edition. Praha : Galén, 2008. 502 pp. ISBN 978-80-7262-371-6.

References

1. POVÝŠIL, Ctibor – ŠTEINER, Ivo – DUŠEK, Pavel, et al. *Speciální patologie*. 2. edition. Praha : Galén, 2007. 430 pp. pp. 394. ISBN 978-807262-494-2.