Bullous and Vesicular Dermatoses

Zaporozhye 2016

Theoretical part

The group of vesicular and bullosus dermatoses includes different diseases on the basis of etiology and pathogenesis (pemphigus, Duhring's dermatosis, simple vesicular lichens, herpes zoster, exudative multimorphic erythema).

True (acantholytic) pemphigus

Pemphigus is a malignant, serious disease. Its clinical manifestation is the formation of vesicles on non-inflamed skin and mucous membranes. If not treated the bulloses soon appear on the whole skin. Patients should consult not only dermatologists but also other specialists (physicians, dentists, infectionists). Due to this, the knowledge of this pathology is necessary for all the clinicians to render qualified help to the patients.

Etiology and pathogenesis of pemphigus

There are different etiopathogenic theories, in particular, the viral theory but it is not completely proved. Recently autoimmune processes are considered to be of great importance in the pathogenesis: discovery of antibodies to intercellular substance in the skin, in liquid of the bulloses and in blood serum. In immunofluorescence, in intercellular space of stratum spinosum of epidermis immunoglobulin G is found only in the patients with pemphigus.

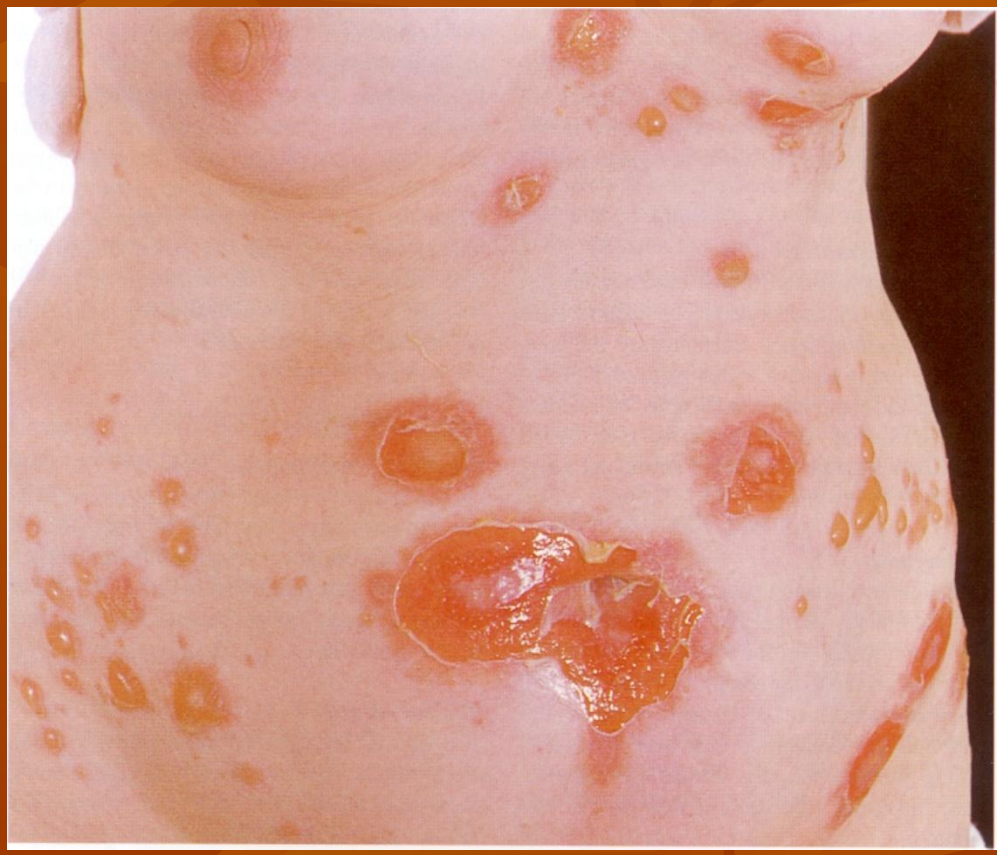
Clinical varieties

Four forms of true pemphigus are differentiated:

pemphigus vulgaris (common)
pemphigus vegetans
pemphigus foliaceus (exfoliative)
seborrheal pemphigus

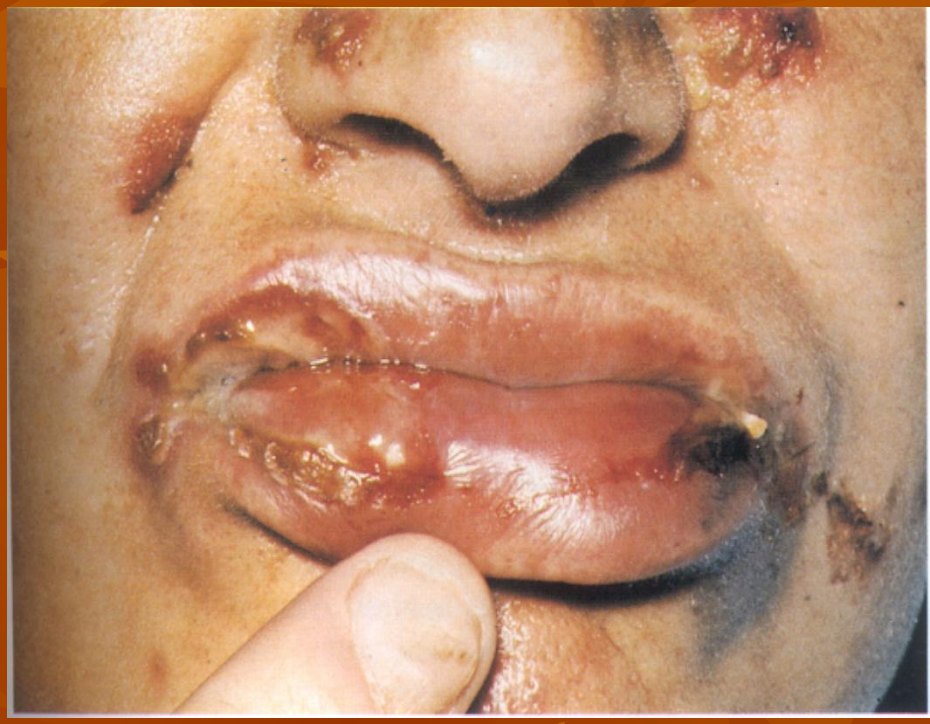
Pemphigus Vulgaris

This form of pemphigus accounts for approximately 75 per cent of the total number of all forms of pemphigus



Pemphigus Vulgaris

Usually dermatosis begins with affection of the oral and throat mucosa, after which, as a rule, the skin of the trunk, limbs, inguinal region, axillae, face and external genitals is involved in the process.

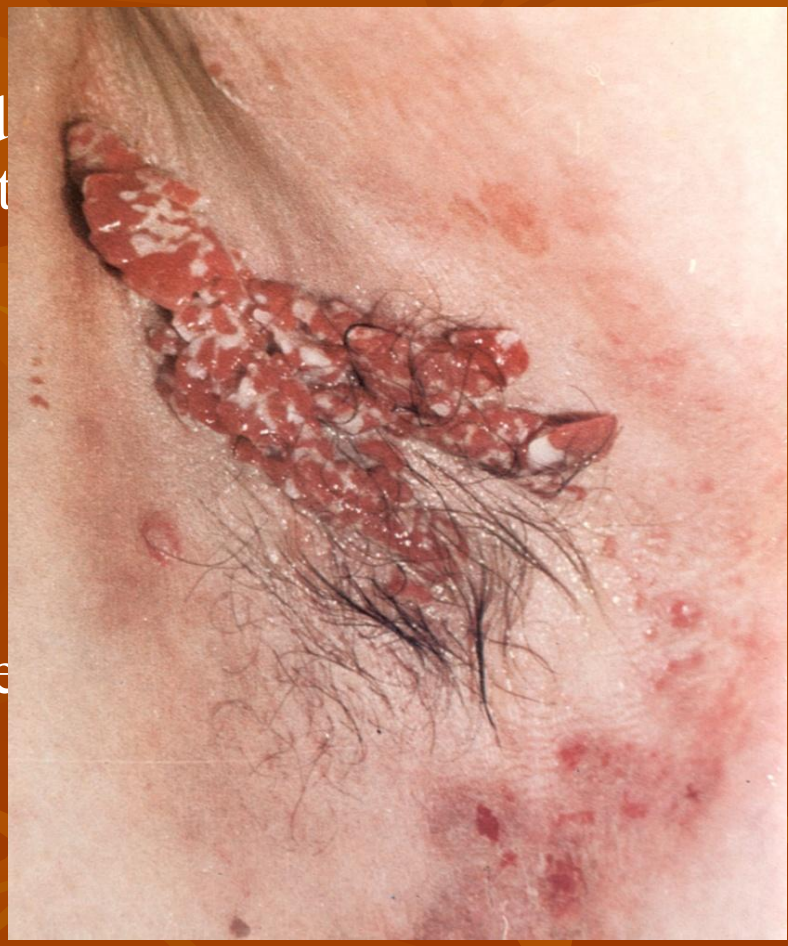


Pemphigus Vegetans

At the beginning of its development this form of pemphigus is clinically similar to pemphigus vulgaris and often starts with the appearance of lesions on the oral mucosa. From the very onset of the disease, however, attention is drawn to the tendency of the bullae to be localized around the natural orifices, the navel and in the region of the large skin folds

Pemphigus Vegetans

Papulomatous growths secreting a considerable amount of exudate are formed later in places of the ruptured bullae against the background of an eroded surface covered with a dirty film. The lesions tend to coalesce and form large vegetative surfaces at places with purulent necrotic disintegration. Nikolsky's sign is often positive. The dermatosis is accompanied with pain and a sensation of burning. Active movements are difficult because of sharp pain



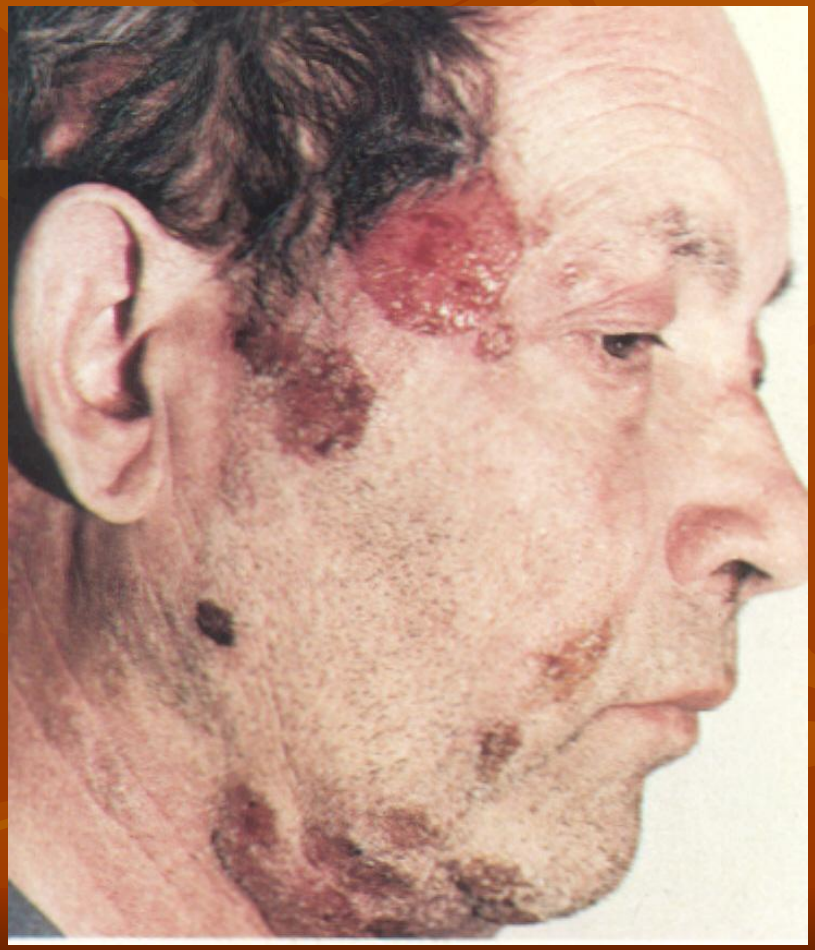
Pemphigus Foliaceus

The disease is characterized by drastic acantholysis leading to the formation of superficial fissures directly under the horny layer, which later turn into bullae.

At the beginning of the disease, flaccid bullae with a thin top and slightly elevated above the surface form on apparently healthy skin. They rupture rapidly with the formation of large erosions. More frequently the tops of the bullae dry up into thin stratified scaly crusts. Epithelization of erosions under the crusts is slow. New portions of the exudate cause the layering of these crusts, producing a scaly surface, hence there is a term 'exfoliative', by which the disease is also known. It is in this variant of pemphigus that the sign described by Nikolsky in 1896 is always sharply positive.

Pemphigus seborrhoicus, or erythematosus (Senear-Usher syndrome)

Pemphigus seborrhoicus belongs to the group of true pemphigus because the possibility of its development into the foliaceus or vulgaris variant has been authentically proved.

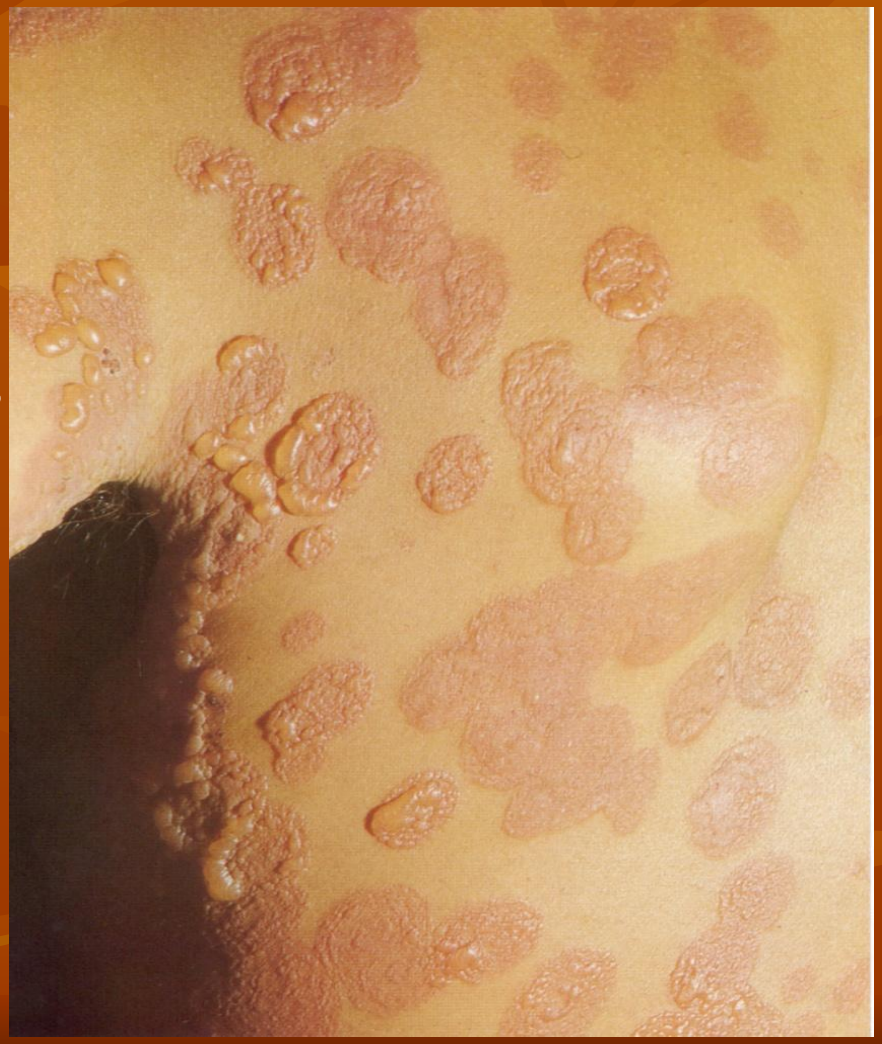


Pemphigus seborrhoicus, or erythematosus

The lesions usually first appear on the face and less frequently on the scalp, chest and back. Closely arranged greasy crusts are formed against an erythematous background and simulate the picture of cicatrizing erythematosis on the face. Moist eroded surfaces are exposed when the crusts are removed. Acantholytic cells are seen in impression smears from these surfaces. The bullae are often formed unnoticeably and the crusts seem to be primary lesions. In other cases vesicles covered with stratified yellowish crusts are formed on the trunk and limbs, in sites characteristic of seborrhea. Lesions rarely occur on the mucous membranes, but if this happens, they are a bad prognostic sign.

Diagnosis and differential diagnosis

Differential diagnosis is carried out with multiform exudative erythema, bullous toxicoderma (Lyell's syndrome) and Duhring's dermatosis

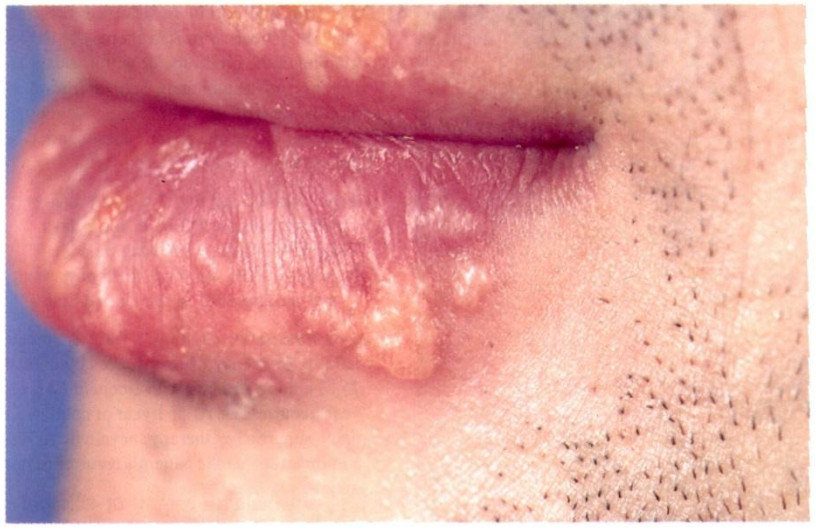


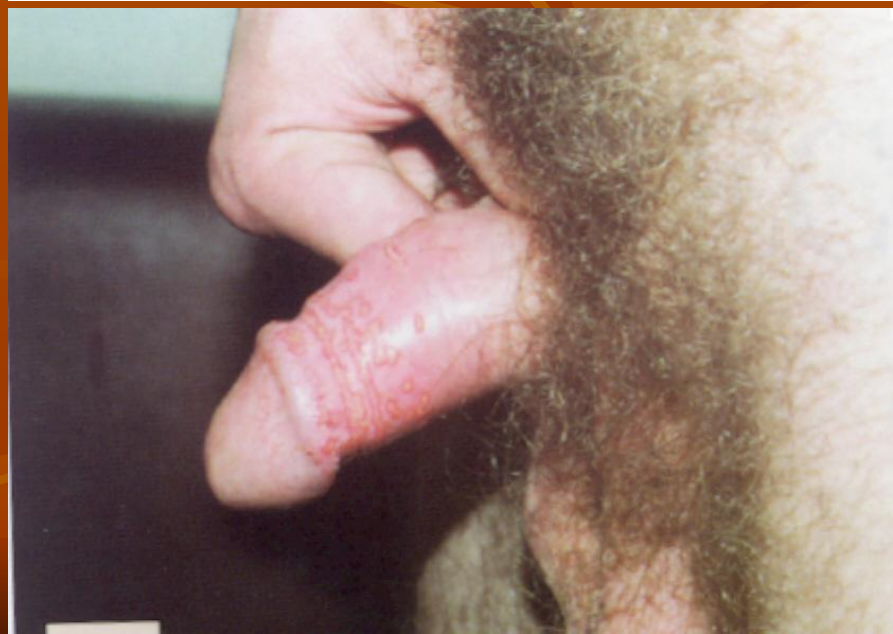
Dermatitis herpetiformis (Duhring's disease)

Differential sign	Acantholytic pemphigus	Dermatitis herpetiformis (Duhring's disease)
Primary elements	Vesicles on the healthy skin	Vesicles, bulla, macula, papule, pustule (polymorphism)
Character of lesions	Flaccid, thin cover soon opens	The cover is dense, exists for 7-10 days
Erosion	Often extensive, without the tendency towards epithelization	Often less; soon heals
Localization of the vesicle	Intraepidermal	Subepidermal
Character of the elements	Monomorphic	Polymorphic
Localization of elements	Unsystematic	Geometric, i.e. grouped
Infection of the mouth cavity	In most patients the disease often starts in the mouth cavity	Very rare; does not start in the mouth cavity.
Nicolsky's symptom	Positive	Negative
Itch	Rare	Usually intense
Course	Continuos course	Relapsing
Prognosis	Serious, in severe cases may be lethal	More favourable, rare transformation into pemphigus
Tzanck acantholytic cells	Discovered	Not discovered
Reaction with iodine	Absent	Causes intensification
Eosinophilia	Not high in blood; rare in vesicles	Frequent in blood (20-30%) Nearly always high in vesicle
Therapeutic effect	Absent	Marked in many cases

Herpes simplex

The disease is caused by a filtrable virus and is characterized by eruption of grouped vesicles filled with a clear and then thick content, on a hyperemic area. The primary sites are the lips (herpes labialis), cheeks (herpes facialis), the wings of the nose (herpes nasalis), oral mucosa (herpes buccalis), cornea (herpes corneae), and genitals (herpes genitalis).





Treatment

- ***Treatment*** with steroid hormones envisages the prescription of maximum doses (80-100 mg of prednisolone or methylprednisolone, 4-6 mg of dexamethasone daily). When new eruptions cease to appear and exudative phenomena in the foci diminishes, the dose of corticosteroids may be reduced, but very slowly (to avoid exacerbation of the process and prevent the 'phenomenon of steroid drug discontinuation').

Treatment.

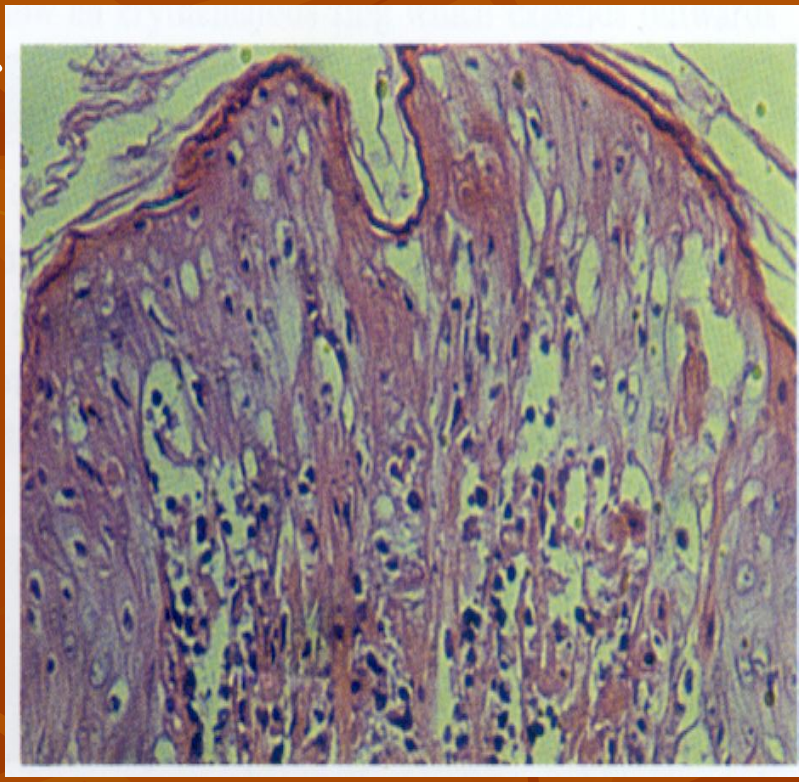
Drying and disinfectant agents are used externally: lotions of Aq. Goulardi and 1-4 per cent silver nitrate solution, 1-2 per cent pyoktanin (gentian violet) solution, 1-2-3 per cent oxolinum or 30-50 per cent interferon ointment, Bonaphton, gossypol, tebrophenum, 1-3 per cent Florenal ointment, and sulfur-carbolic paste (Ac. carbolici 1.0, Sulfuris 1.5, Pastae Zinci 30.0).

The following clinical forms are distinguished:

- mild; with rapid resolution of the few lesions that have erupted;
- edematous; accompanied with bright hyperemia and marked swelling;
- severe;
- zosteriform;
- frequently recurring; localized on the lips, buttocks, and external genitals.

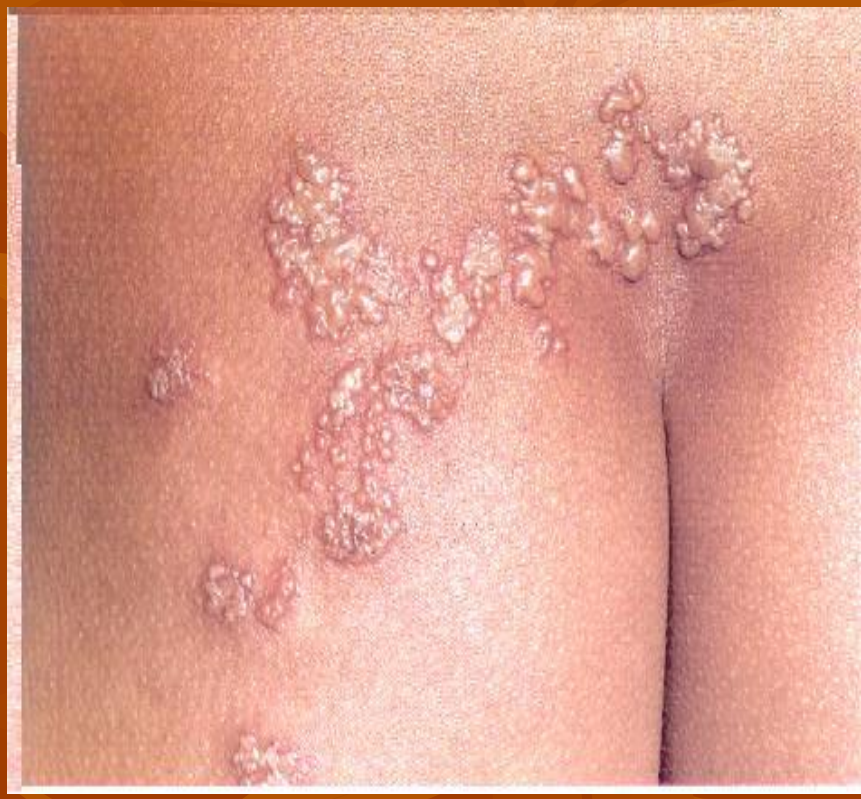
Histopathology

The characteristic findings are ballooning and, to a lesser extent, reticular degeneration of the epidermal cells and acantholysis. There are intranuclear eosinophilic inclusions in the ballooning cells and dilated blood vessels, edema, and mild perivascular infiltration in the dermal papillary layer.



Herpes zoster

Herpes zoster, also called shingles or zona, is caused by a neurotropic filtrable virus Strongiloplasma zonae which resembles or is identical with the chickenpox virus in antigenic structure and the ability to grow in human embryonic tissues



The following clinical varieties are distinguished:

- generalized herpes zoster marked by bilateral and disseminated lesions;
- herpes zoster haemorrhagicus, in which the clear contents of the vesicles turn purulent and then, when the process penetrates deeper into the dermis, become hemorrhagic;
- herpes zoster gangraenosus, a severe form, in which the floor of the vesicles undergoes necrosis and scars are formed in their place;
- mild form;
- bullous form characterized by the appearance of both vesicles and bullae.

Histopathology.

Ballooning and reticular degeneration of the epidermal cells, intranuclear viral inclusions, and degenerative changes in the nerve fibres may be seen. Acute inflammatory polymorphonuclear infiltration predominantly of a lymphocyto-histio-cytic character, edema, and dilated blood and lymph vessels are revealed.

Treatment.

Antiviral drugs such as methisazonum or cutizonum (one taken two or three times a day for three to six days), interferon, salicylates and analgesics (acetyl salicylic acid, amidopyrine, phenylbutazone, rheopyrine), vitamins B1, B5 B6, B12, and C, autohaemotherapy, gamma globulin injections, and interferonogens are prescribed.