IVANOVA I.N.

DISEASES OF CONNECTIVE TISSUE. LICHEN RUBER PLANUS.PSORIASIS.

- Definition: the group of poligenic, multifactorial diseases with alteration of connective tissue, systemic disturbance, chronic duration.
- Clinical picture depends on genetic predisposition.
- Patients with genetic predisposition for systemic duration have alteration of internal organs and systems contrary to patients with skin forms.

Classification

1. Lupus erythematodes.
2. Sclerodermia.
3. Dermatomyositis.
4. Periarteriitis nodularis.

Lupus erythematodes

- **Lupus erythematodes –** collagenosis characterized by inflamation of skin, mucosae, kidneys, hart, liver, joints, blood vessels.
- **Etiology. Pathogenesis.**
- Disease is poligenic, multifactorial. May be dominant, recessive genetic line.
- The manifestation, onset may be at any age of patients.
- Histological picture:
- 1)hyperkeratosis at the ostium of hair follicle;
- 2)vacual dystrophy of basal layer of epidermis;
- 3)lymphocitar infiltration of dermis;
- 4)vasodilatation in superficial part of dermis.

ETIOLOGY AND PATOGENESIS.

- The general antigen in Lupus Erythematodes is dischanged DNA of leucocytes, witch results in synthesis of Ig G and immune complexes. The application of this complexes depends on genetic predisposition: basement membrane, papillary layer of dermis, endocard, myocard, pericard, kidneys, joints, mucosae. Skin chronic forms of lupus erythematodes never transform into systemic.
- Alteration of connective tissue in duration pass trough 3 stages:
- 1)mucoid oedema;
- 2)fibrinoid necrosis;
- 3)sclerosis (atrophy).

ETIOLOGY AND PATHOGENESIS.

If the immune response caused by dischange of DNA, all mutagen factors participate in pathogenesis of LE:

1)different rays (ultraviolet, X-ray, sunlight);

2)drugs (antibiotics, first of all penicillin) and others;

3)Infections (topic, acute, vaccinations);

4)professional unhealthy;

5)in women – level of estrogens (pregnancy, delivery).

CLASSIFICATION

1. Discoid.
2. Disseminated.
3. Centrofused (usually systemic).
4. Deep (Kaposy-Irgang).
5. Systemic (acute, subacute, chronic).

CLINICAL PICTIRE

- The typical location of lupus erythematodes is face and open parts of skin.
- Skin rash is characterized by:
- 1) vascular spot of pink, cyanotic color , sharply demarked, infiltrated;
- 2)hyperkeratosis at the place of hair follicle (“sharp ladies heel symptom”, Meschersky symptom”).

3) cicatricial atrophy (depigmentation, thin skin, absence of hair, teleangiectasia).

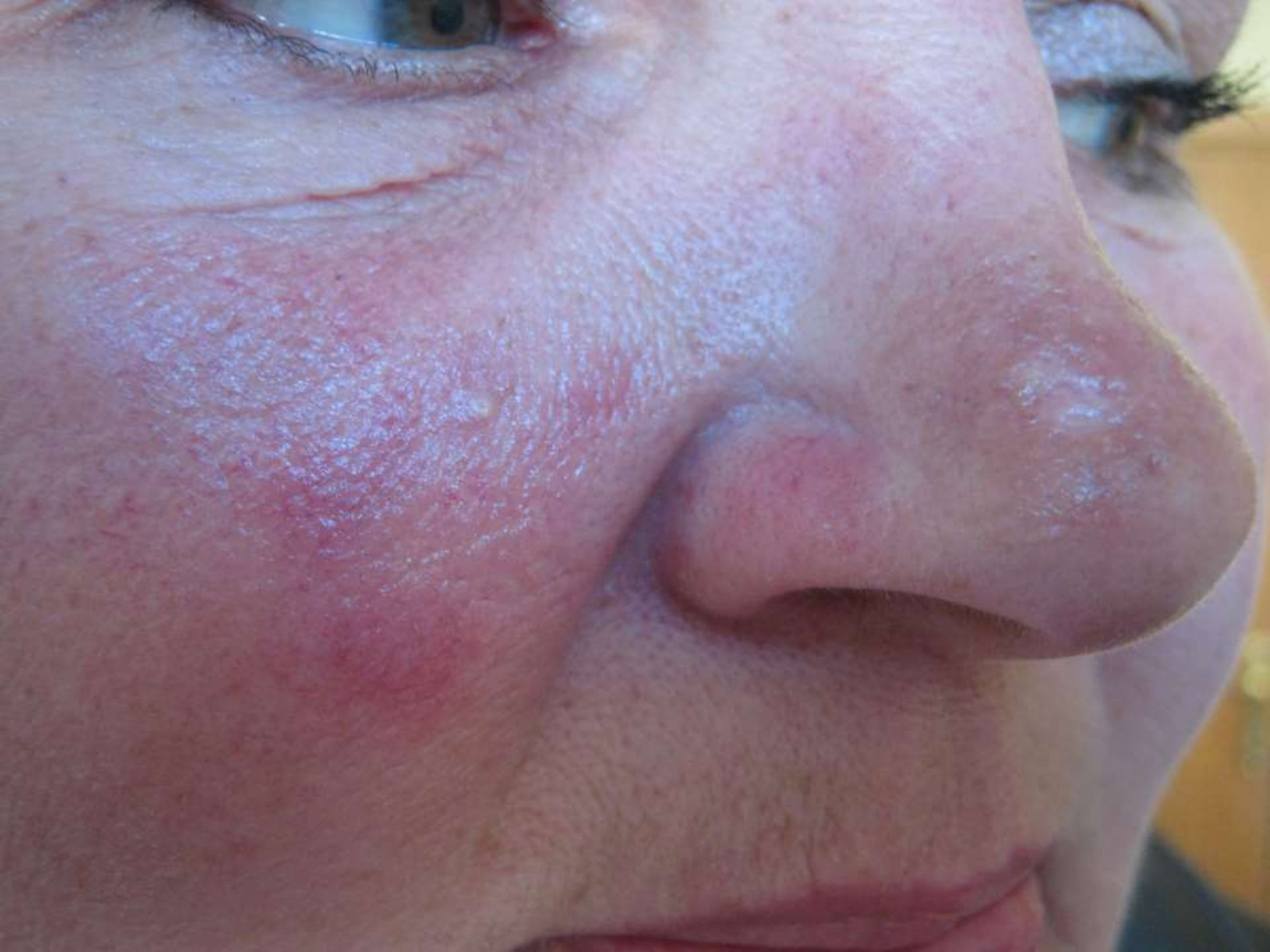
- **Disseminated lupus erythematodes** is characterized by location of leasons on all parts of the skin, not only on the head.
- In oral cavity LE may be on cheeks, lips, tong. Atrophy of this organ results in erosions, patch, hyperkeratosis.

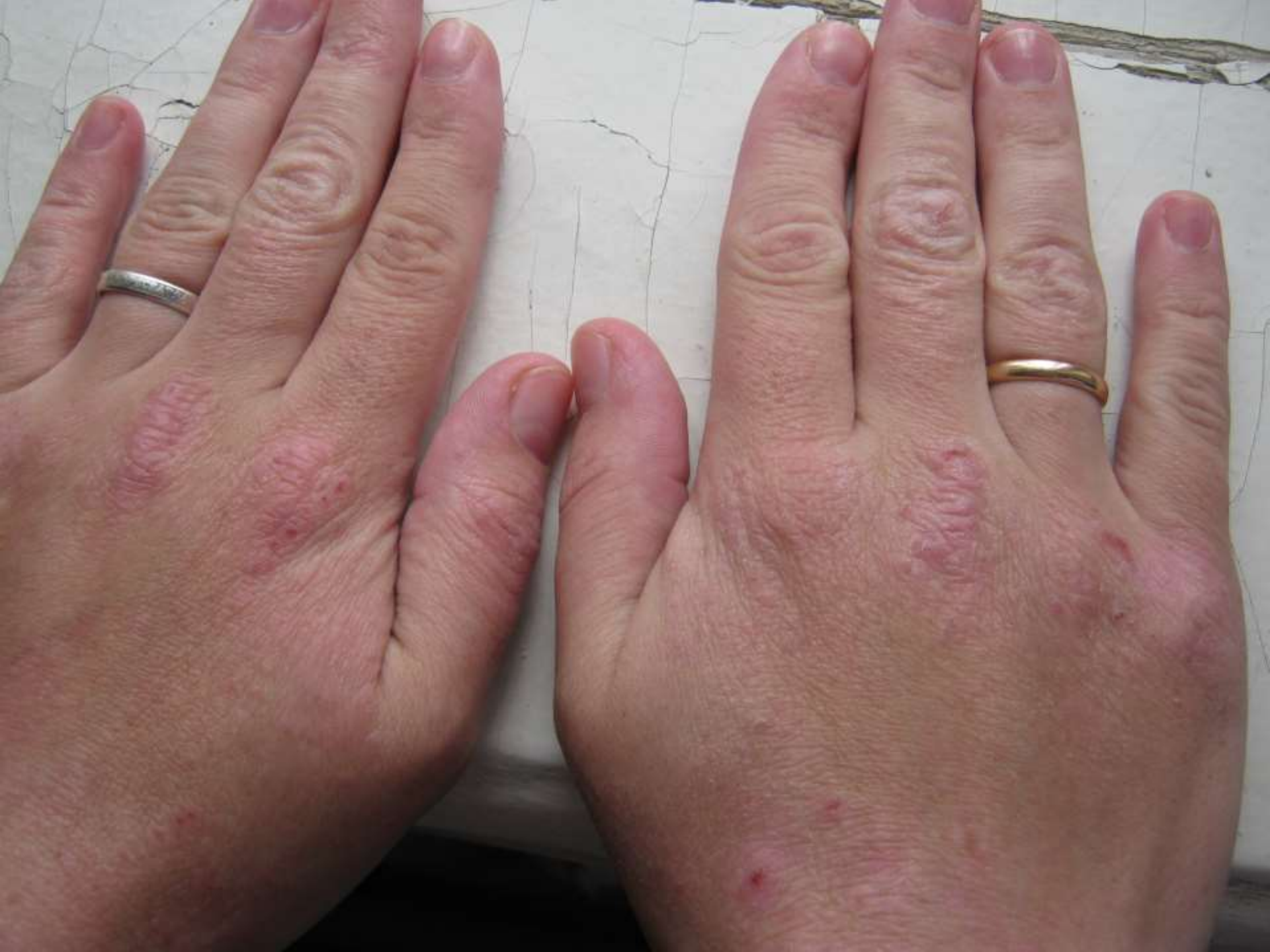
CLINICAL PICTURE

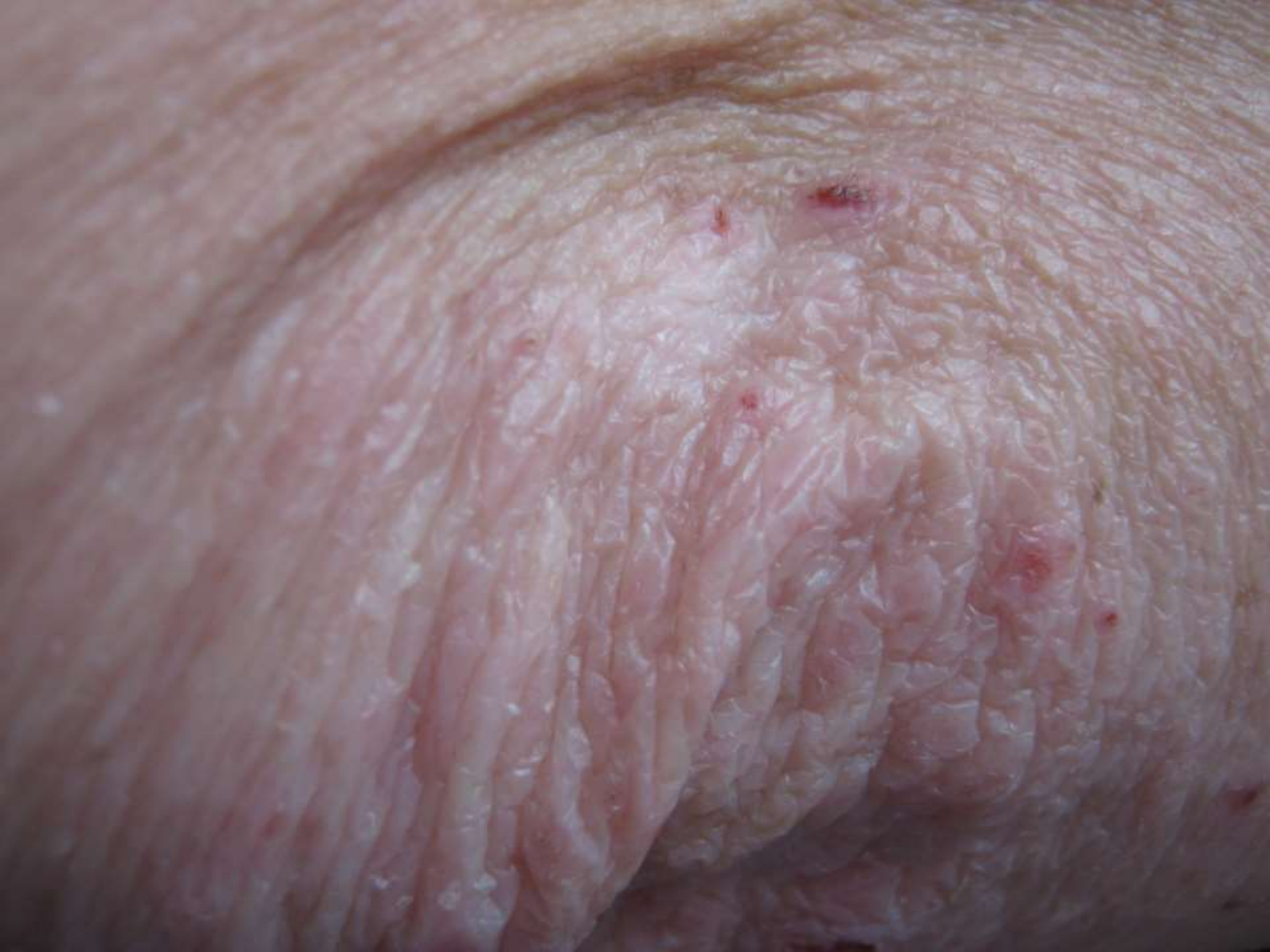
- **Systemic lupus erythematodes** includes alteration of inner organs hard duration. Its typical to see high fever, bad condition, asthenic syndrome, distal vasculites, arthropathia, Rejno syndrome, hepatosplenomegalia.
- **Laboratory signs:** anemia, leucopenia, trombocytopenia, high sedimentation of erythrocytes in blood analysis. The urine analysis is typical for glomerulonefritis: proteinuria, cylindruria, dischanged erythrocytes.
  Electrocardoigramma may be typical for endo-, myo-, pericarditis.
- X-ray examination shows specific abnormality of lungs – pneumonitis.
- In patients without any leasons on the skin disease is called lupus without lupus. Verification includes only laboratory and instrumental examinations.

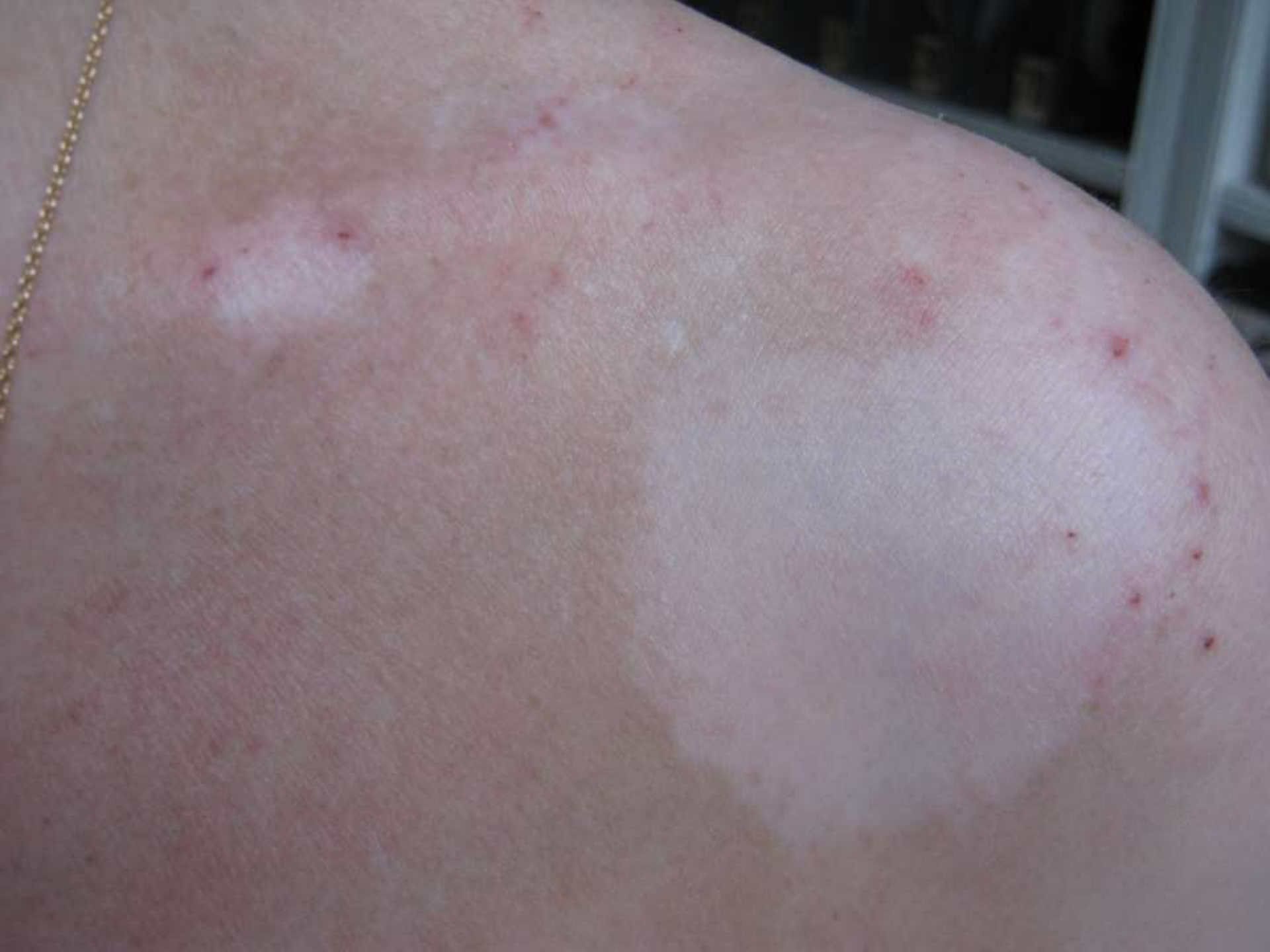
CLINICAL PICTURE

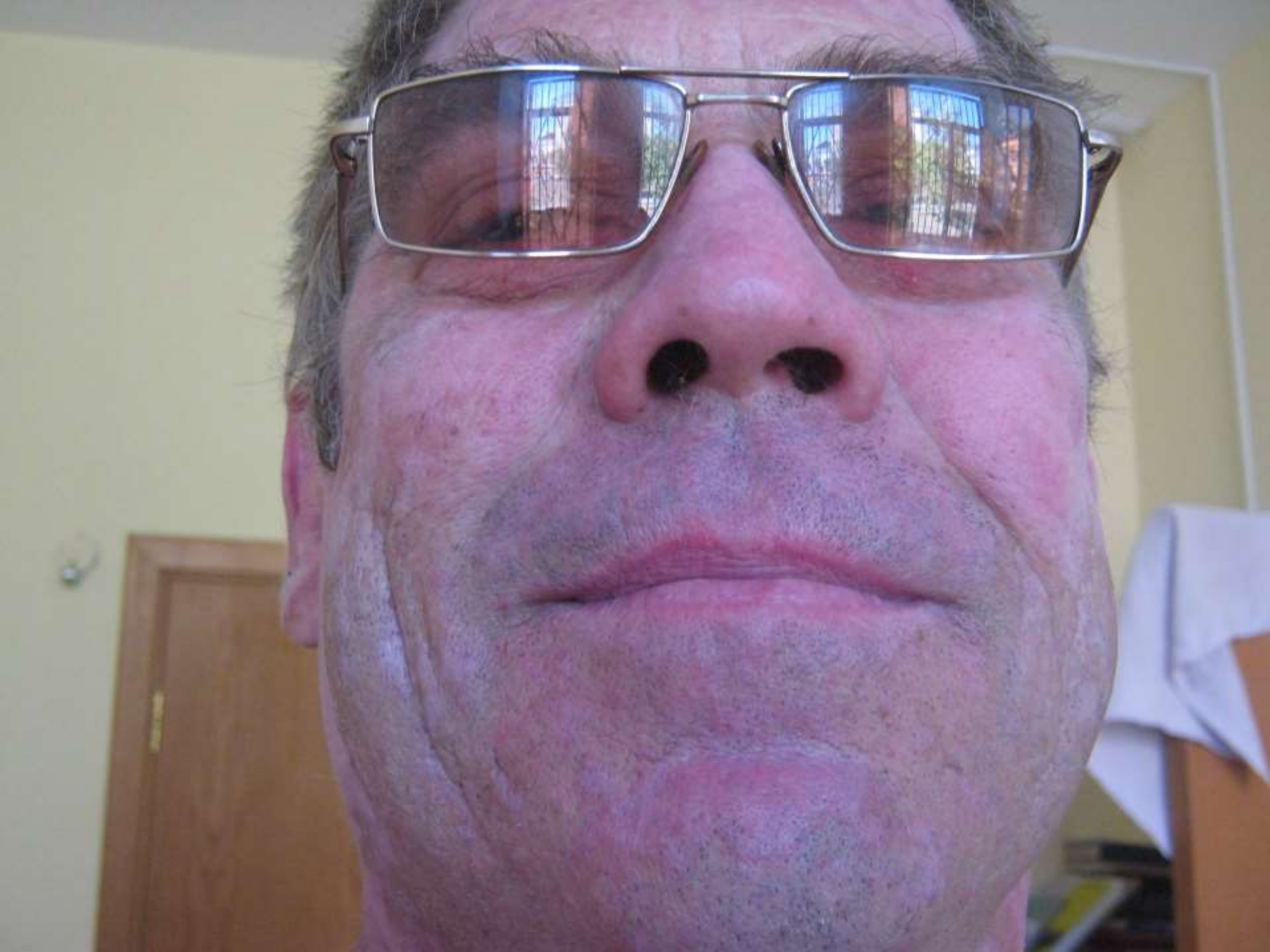
- Biett erythema centrifugum is systemic form of lupus erithematodes with superficial alteration of face and absence of cicatricial atrophy.Biett form is typical for children and pubertate patients. Different internal signs of LE may be: lupus endo-, mio-, pericarditis, lupus glomerulonefritis, polyartritis, Reino syndrome.
- Kaposi – Irgang lupus erythematodes is systemic profound form of LE with inflammation of hyperdermis (panniculitis). Clinical picture is characterized by nodes, ulcers and scars. May be only skin forms.

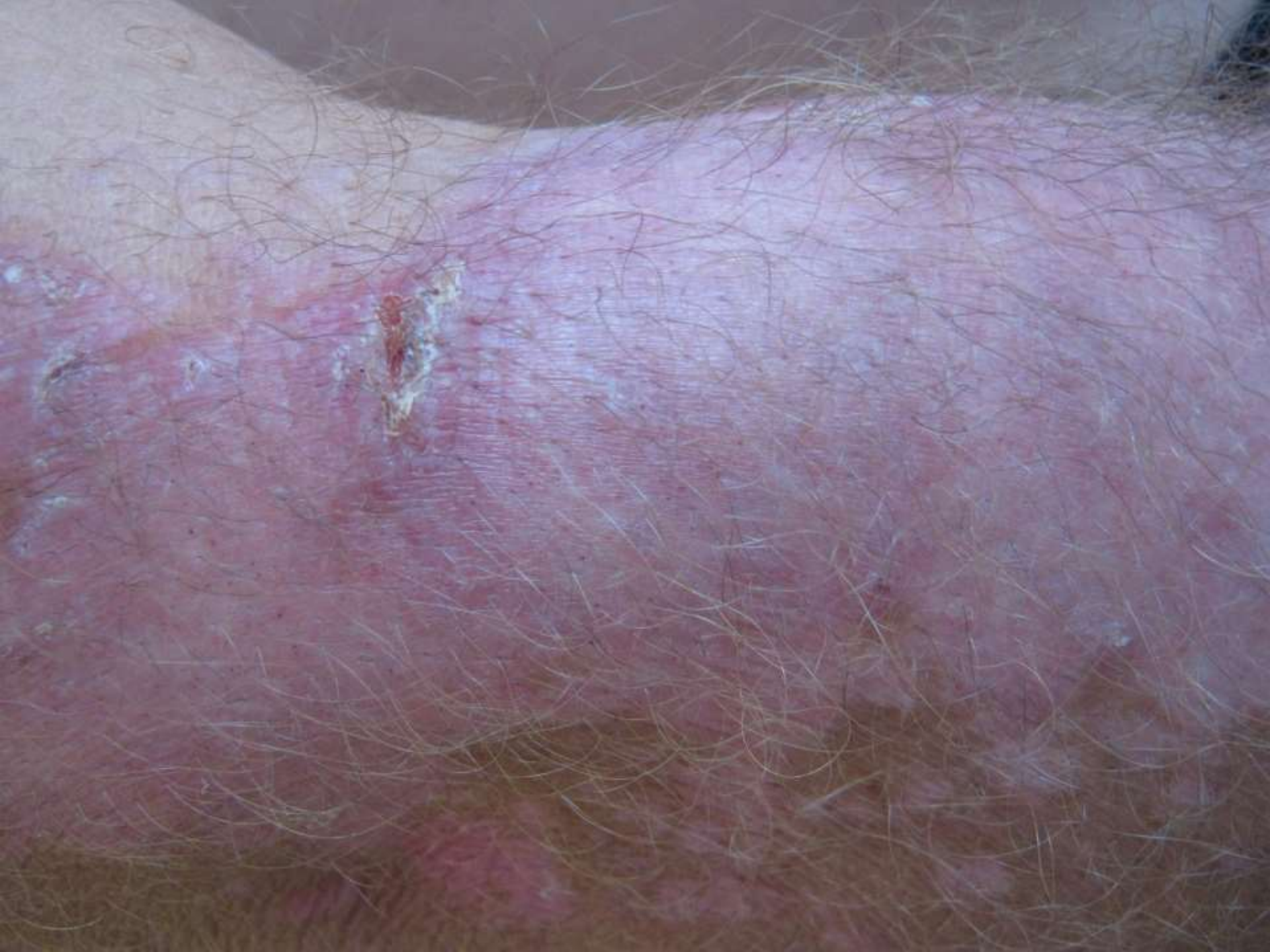












TREATMENT.

The treatment of discoid lupus erythematodes includes:

1) management of antimalarian drugs (delagil, resochin, plakvenil) in sprin (0,25 mg 3time a day during 10 days, after 0,25 a day during 50 days, in children the half doses are used; complications: leucopenia, retinopathia;
2) vitaminotherapy (vitamins B group, B6, B12, B 15), vitamin A natural and retinoids (aevit, radevit, neotigason);
3) in disseminated form of LE may be used small doses of common corticosteroids (metipred, prednisolon) 30-40 mg and cytostatic asatioprin 1-2 mg on kg;
4) Phonophoresis of ungventum hydrocortisoni 1% on lesions of LE, topic corticosteroids without fluorine.

TREATMENT

- Treatment of acute and subacute forms of LE includes stroke doses of prednisolon (100mg, 20 tab. a day). After breacfast 10 tab., after dinner – 7 tab., after supper – 3 tab. Maximum affect is from 6 till 8 hours in the morning. 7-10 days later corticosteroid dose deminisched during long time to minimum affective dose, usually 3 tab.
- This dose, sometimes may be 2-1 tab. Patient must use minimum affective dose during all his life.

COMPLICATIONS

Complcations of common corticosteroid treatment:

- 1) abnormality of protein metabolism (hypoproteinemia, , catabolic affect, aquired immune deficiency, steroid ulcer of stomach, dystrophy of muscles and bones;
- 3) disturbance of water-salt metabolism (potassium deficiency, accumulation of natrium, hypertension, edema);
- 4) abnormality of lipid metabolism (Icenko-Kusching syndrome);
- 5) steroid acne;
- 6) steroid psychosis;
- 7) disseminated intravascular coagulation syndrome, tromboses of life important arteries.

TREATMENT OF COMPLICATIONS

- 1)anabolic steroids (retabolili, calcii orotatis);
- 2)diet 9 (ant diabetic);
- 3)potessium drugs (asparcam, panangin).
- All patients must be on supervision of dermatovenerologist and therapeutist (revmatologist).
- Prognosis is individual and depends on degree of complications.

SCLERODERMA

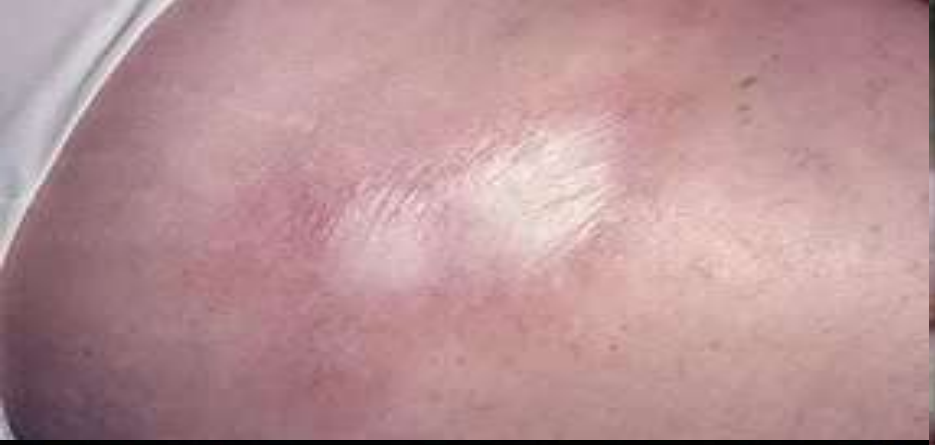
- **Scleroderma –** autoimmune poligenic collagenisis with alteration of collagen fibres and hardening, induration of skin.
- In patogenesis of scleroderma topic infections, borreliosis, mechanical traumas, endocrine disturbanses, surgical treatment, overcooling participate.
- General autoantigen in scleroderma is collagen. Synthesis of IgG results in 3 stages:
- 1) stage of edema (erythema of cianotic color, cold edema);
- 2) stage of hardening, sclerosis (skin is hard, yellow, ivory);
- 3) stage of cicatricial atrophy (thin skin as "cigarette tissue-paper").
- In basis of edema alteration of blood vessels, of sclerosis – deficiency of hyaluronidases.

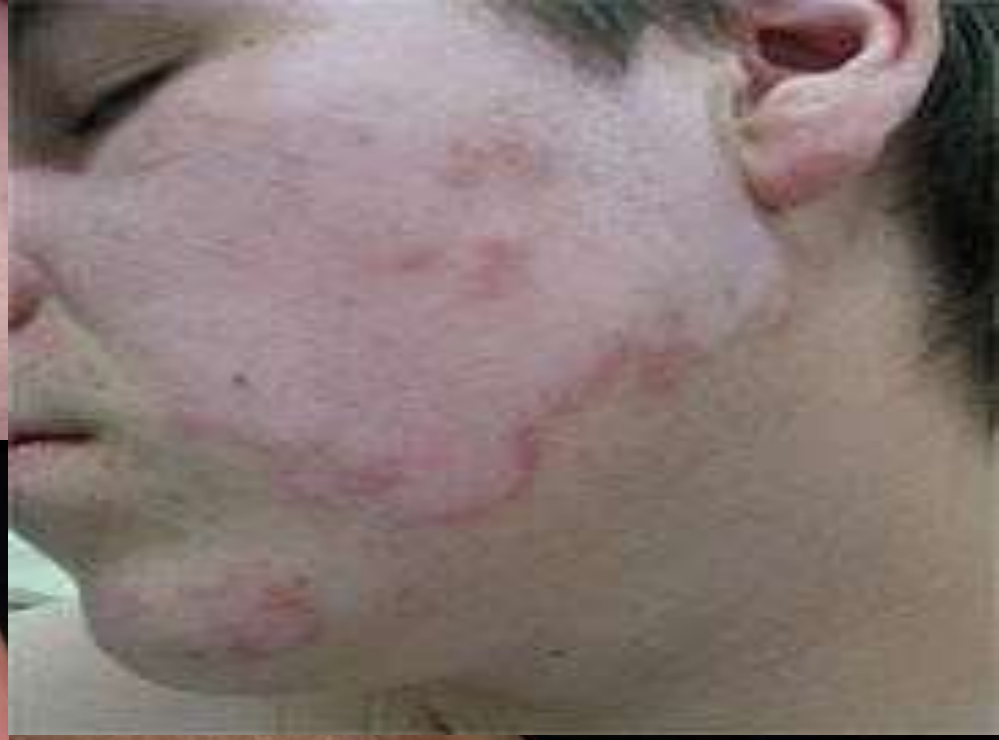
CLASSIFICATION

1. Lesion form scleroderma : plaque form, stripe form. Disease of white macules in some classifications is atrophy of skin, but in other is superficial form of scleroderma. .

2.Systemic form scleroderma: acrosclerosis, diffuse form, CREST syndrome.

- **Plaque form** usually localized on trunk, limbs, is benign form of scleroderma with chronic duration.
- **Stripe form** localized on face, limbs. This form has not tendency to peripheral growing, but to deep growing. That's why typical complications are neurological disturbances, asymmetric face and limbs.
- **Diffuse scleroderma** is charaterized by hardening, yellow color of all skin, mask face, small mouth, thin nose, depigmented hair, dystrophy of hands (birds paws), stenosis of esophagus , alteration of hart, lungs, kidneys.
- **Acrosclerosis** starts with hardening of distal parts of arms, but then is transformed in diffuse scleroderma.
- **CREST** syndrome is characterized by abnormality of calcium metabolism, application in skin: 1)calcinosis, 2)Rejno syndrome, 3)esophagitis, 4)sclerosis of skin, teleangiactasies.



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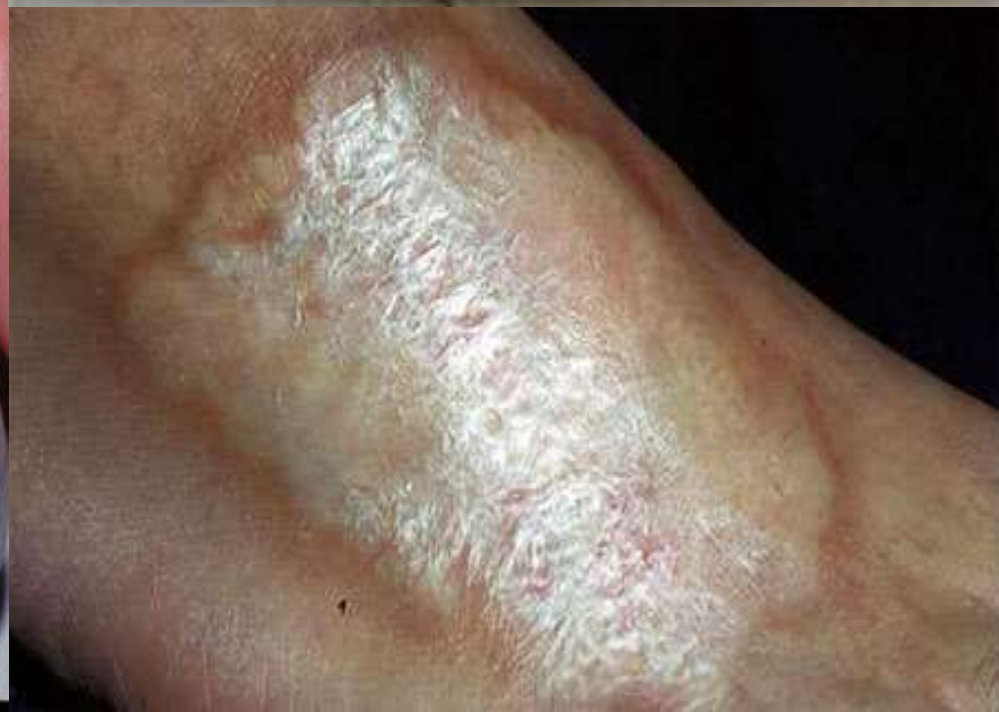


TREATMENT.

- Treatment of scleroderma:
- 1)etiological - cure of topic infections, patholody of GET, treatment of endocrine disturbances;
- 2)pathogenic – according to stage of scleroderma;
- 3)symptomatic – removement of itch or pain.
- In stage of edema is used penicillin, angioprotectors, aloe, placenta extract, fonoforesis of hydrocortison ointment, laser treatment, dimethylsulphoxid.
- In stage of sclerosis must be penicillin, drugs oh hyaluronidase (lidase, longidase), fonoforesis of hydrocortison ointment.
- In stage of athrophy treatment is not necessary except cure of somatic diseases.

TREATMENT

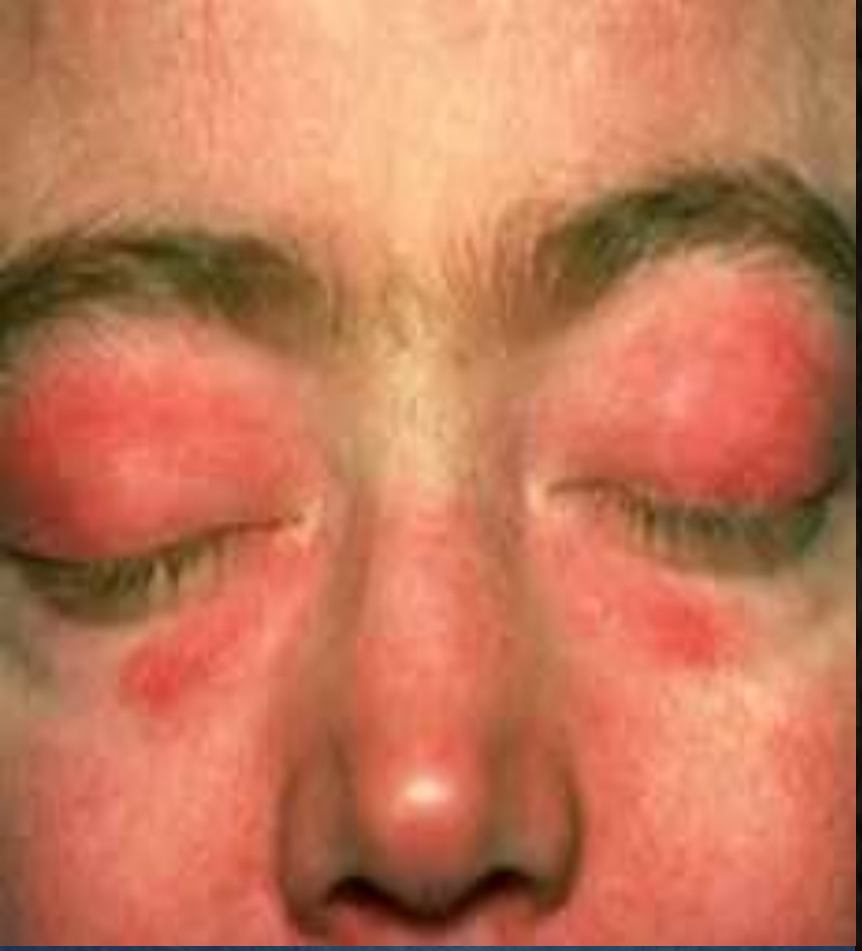
- Treatment of systemic scleroderma is the same as treatment of systemic lupus erythematodes.
- The order:
- 1)stroke doses;
- 2)minimum affective doses;
- 3)supervision during all life.

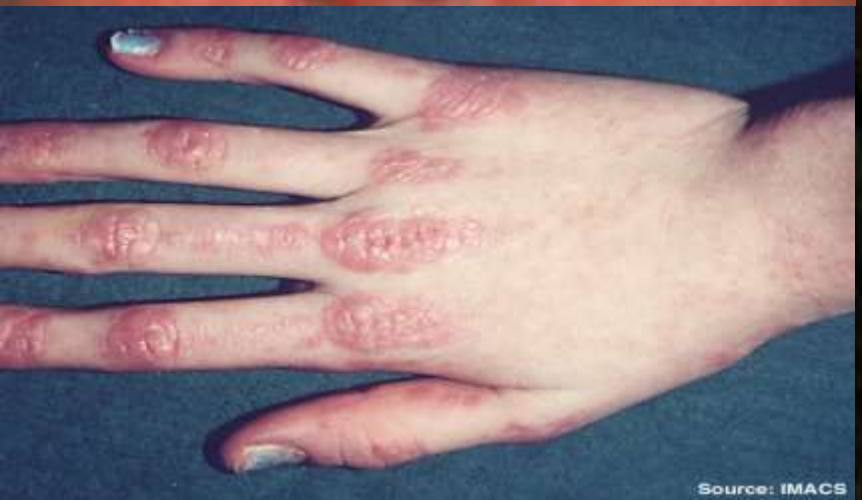
DERMATOMYOSITIS

- **Dermatomyositis** 1s collagenosis with synthesis of auto-antibodies to mussel tissue , usually paraneoplastic etiology in adults and benign duration in children.

CLINICAL PICTURE

- **Dermatomyositis** is typical for women. On face around eyes (periorbitalis location) erythema with lilac color occurs ("glasses symptom"). In mussels must be weakness. On distal parts of limbs typical vasculitis, Rejno syndrome. May be alteration of hart, kidneys, joints. In children is typical to see recovering before puberties age.

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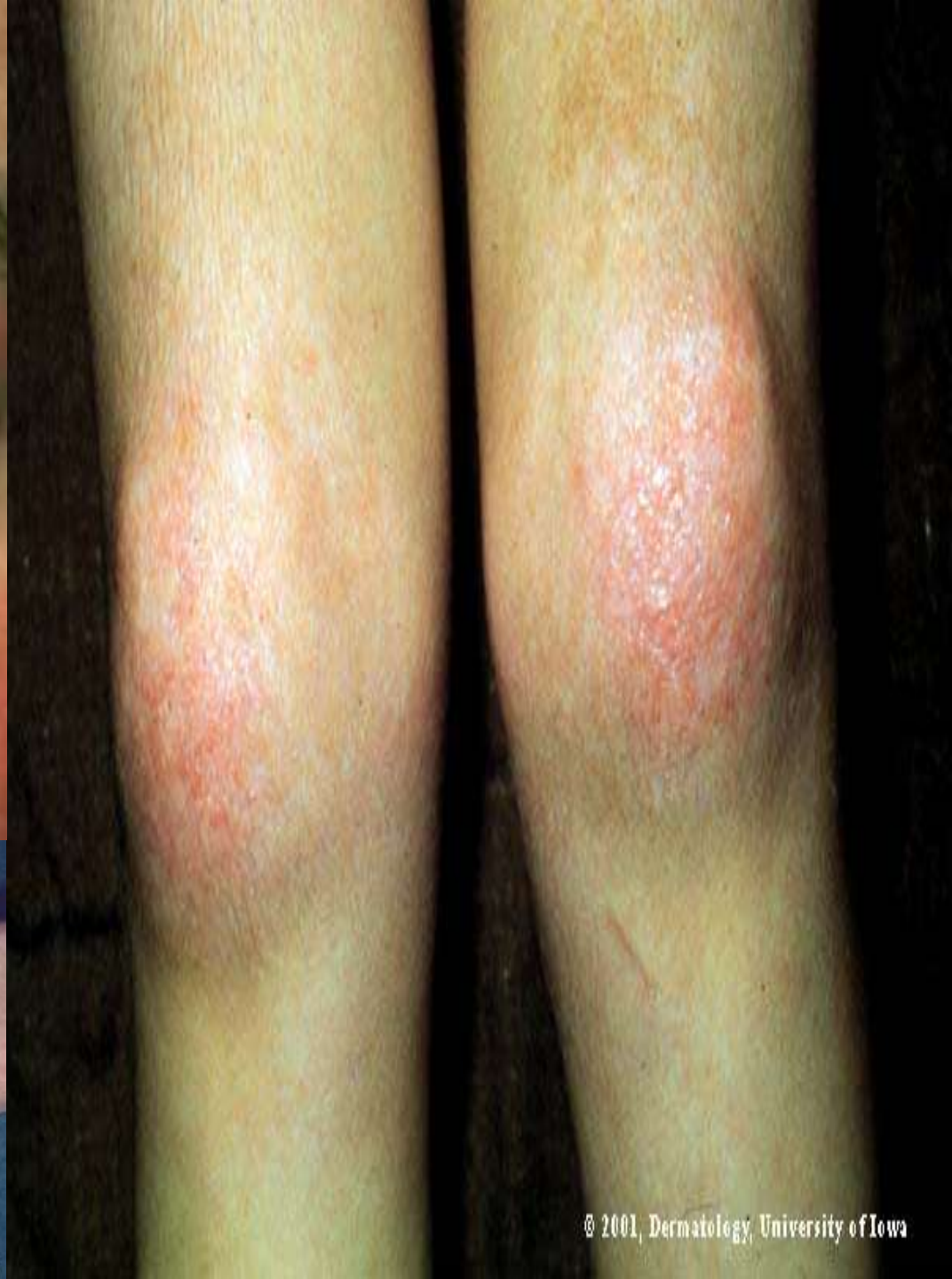
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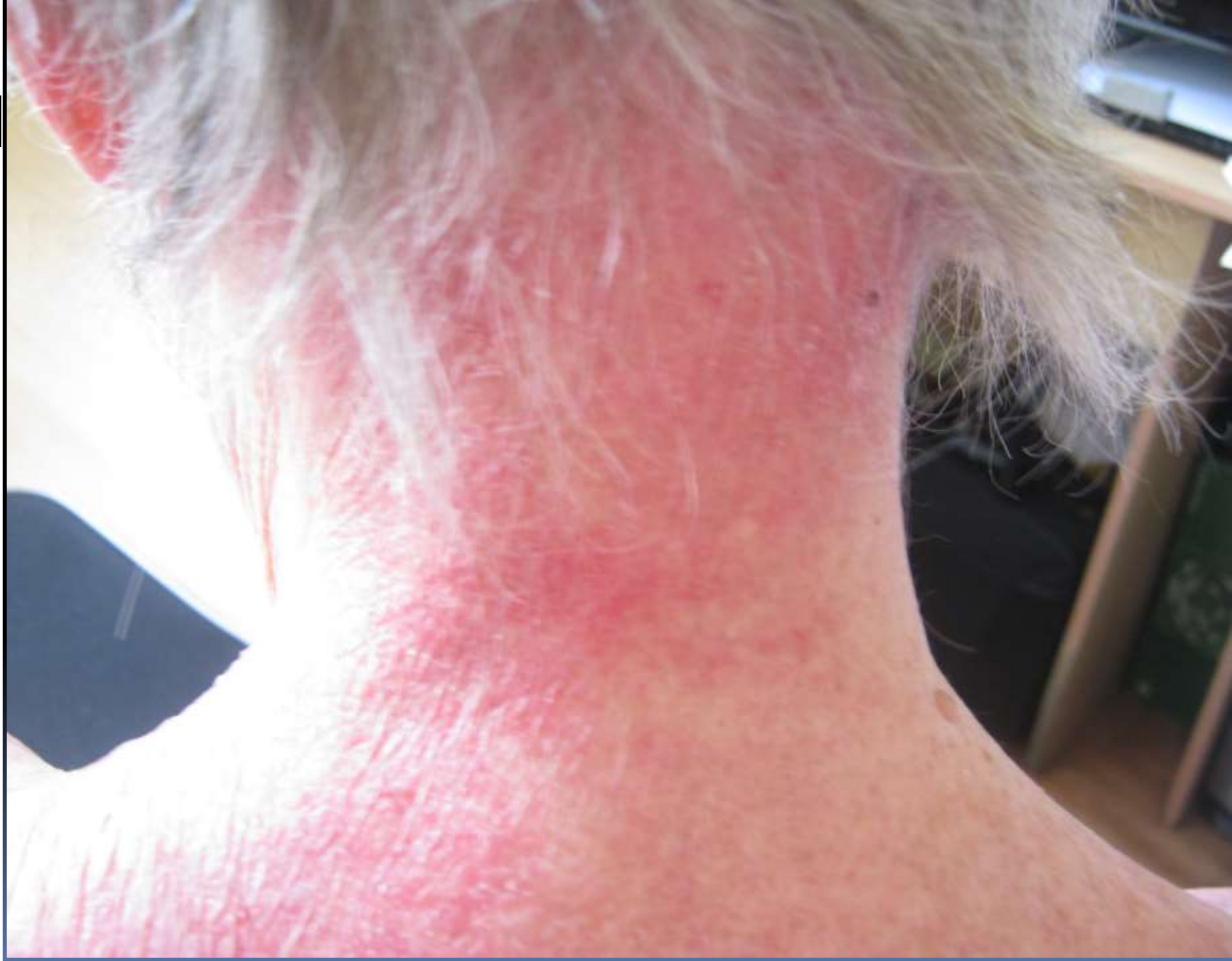
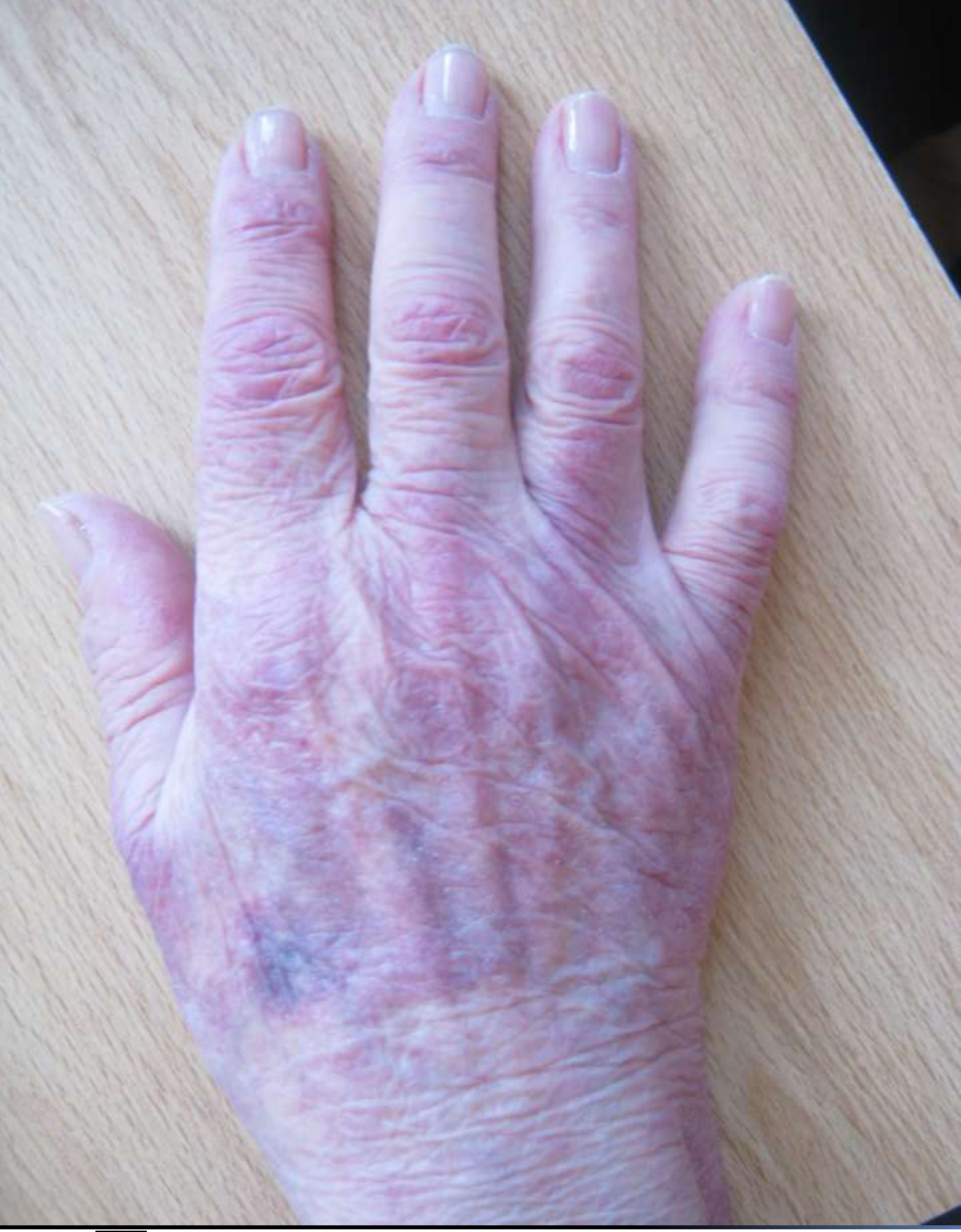
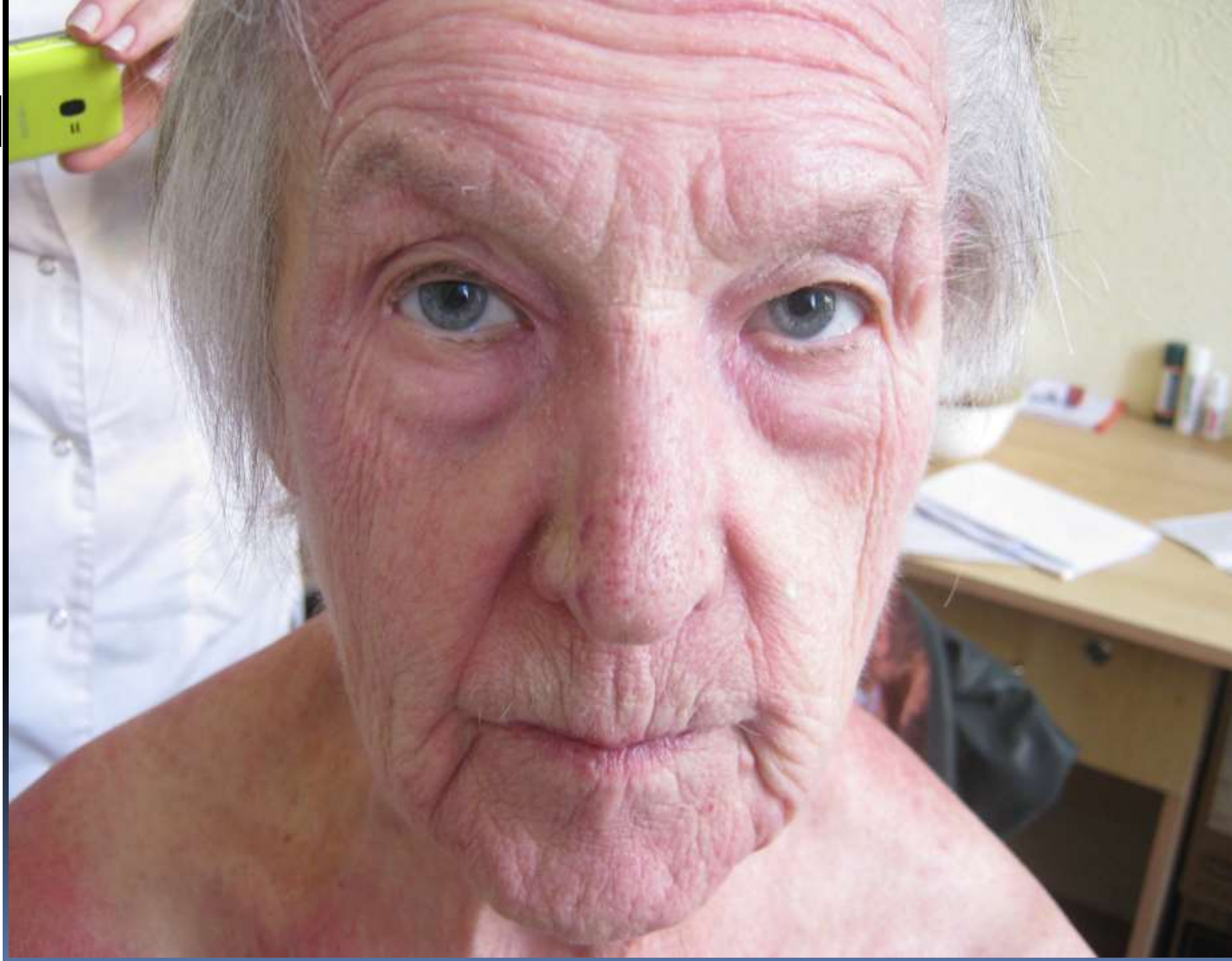






TREATMENT

- 1)etiologic (cure of oncologic diseases);
- 2)common corticosteroids as in treatment of lupus erythematodes and scleroderma;
- 3)cure of complications of steroid therapy.

PERIARTERIITIS NODOSUM

- **Periarteriitis nodosum** is collagenosis with autoantibodies to intima of blood vessels, benign in women and malignant in mails.
- **Clinical picture:** primary elements – papules and nodes cianotic-pinc color localized at the place of blood vessels. Somatic signs the same as in all collagenoses, in mails is typical nephritis, kidneys deficiency.
- Location of lesions in oral cavity may results in destruction of hard palate. This form of periarteriitis nodosum is called Vegener syndrome.

TREATMENT

- Treatment of periarteriitis nodosum depends on clinical form.
- In skin form antiinflamative cure is used.
- In systemic forms common corticosteroids in same doses as in other collagenoses are managed.
- Symptomatic treatment includes angioprotectors, diuretics, topic drugs.

LICHEN RUBER PLANUS

- LRP is chronic disease with itch or pain papules on skin, mucosae, external genitalias, of recurrent duration.
- Mostly LRP is psychosomatic disease in form of masked depression.
- LRP may associated with diabetes, abnormalities of GET.
- Hysthological picture includes vacuole degeneration, acanthosis, granulosis, hypergranulosis in epidermis, lymphocytar infiltrate in dermis.

CLASSIFICATION

1. Classic LRP is characterized by flat papules pink or lilac color on typical location (flexors of limbs, wrists, small of back, front surface of shines, oral mucosa. The surface of papules is shining with white network (Wikhem syndrome). Secondary elements are hyperpigmented macules, seldom depigmented athrophic lesions.
2. Hypertrophic (verrucose) LRP is form with thik horny layer on surface of primary elements and typical location on surface of shines.
3. Athrophic LRP is characterized by thin skin, depigmentation, cicatricial athrophy.

CLASSIFICATION

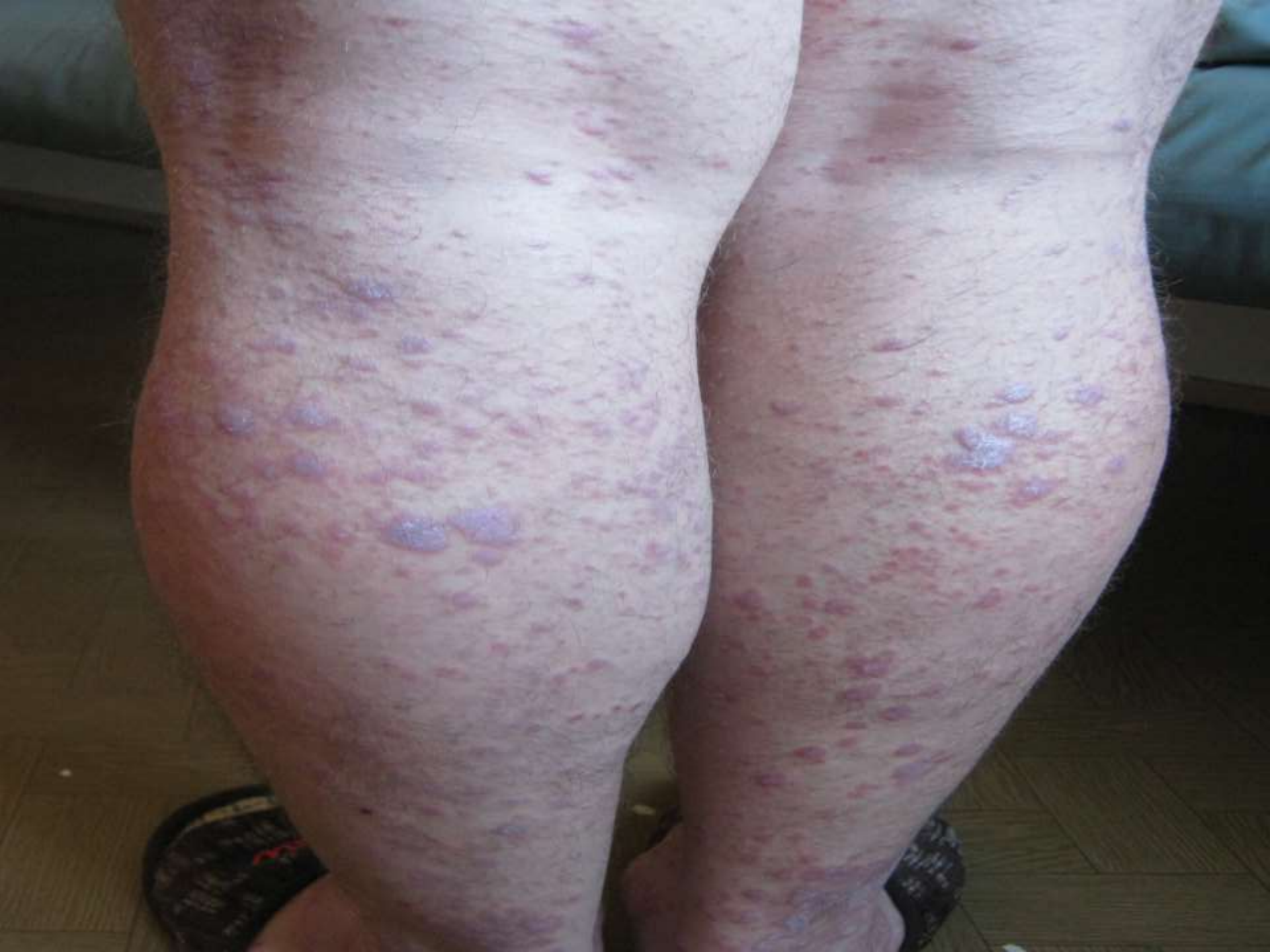
- 4. Blister form is characterized by blisters on surface of papules typical for lichen ruber planus. Secondary elements are crusts with hemorrhagic ecxudate. Clinical picture is similar to clinical picture of During disease.
- 5.Pigmented form usually is paraneoplastic. Papules of LRP are flat, brown, hyperpigmented, localized on big folds, anogenital area. Duration of this form is chronic, reccurent.
- 6. Ring form is characterized by elements of ring form pink color with shiny surface.

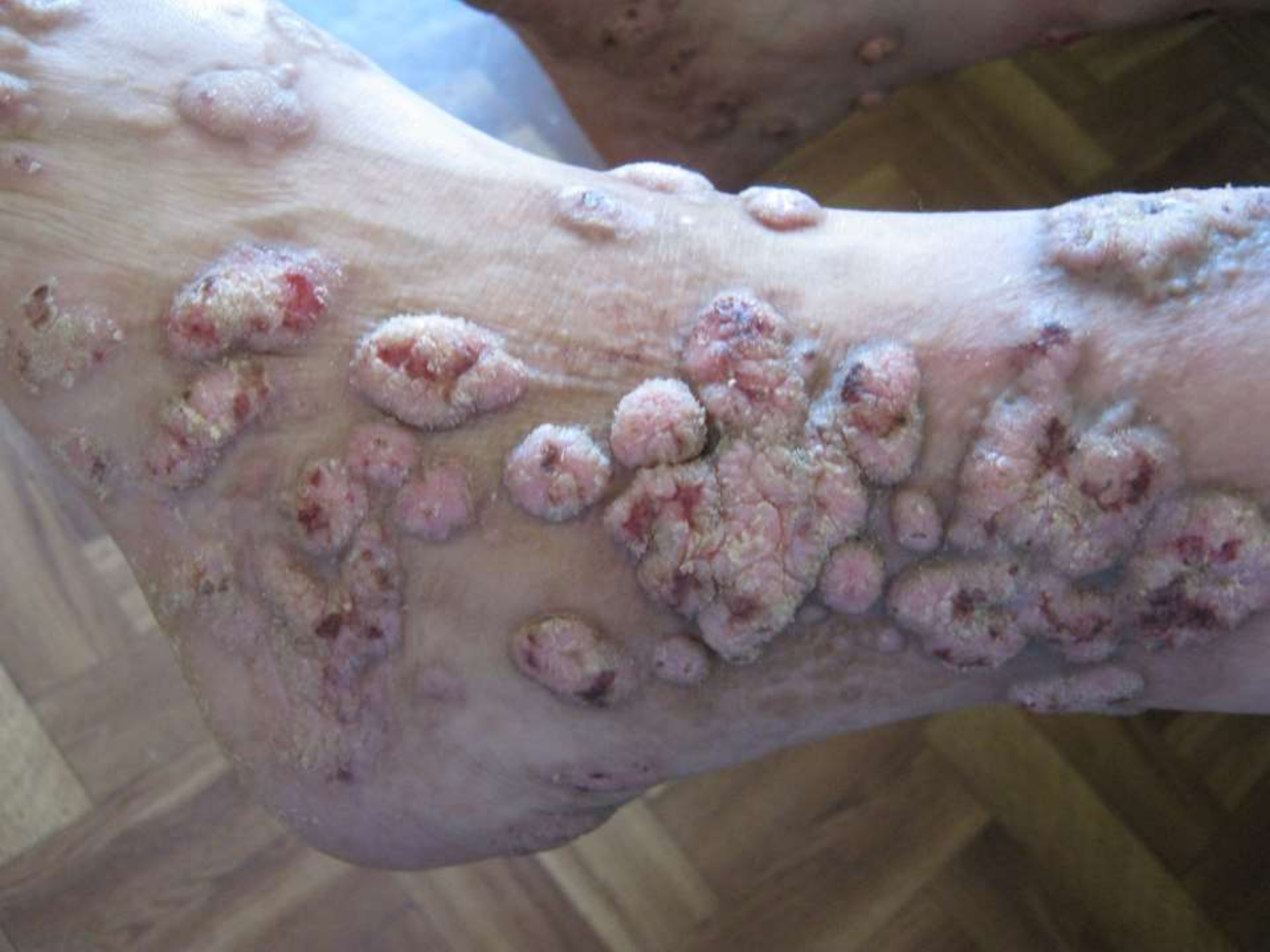
LRP of oral cavity includes 5 variants:

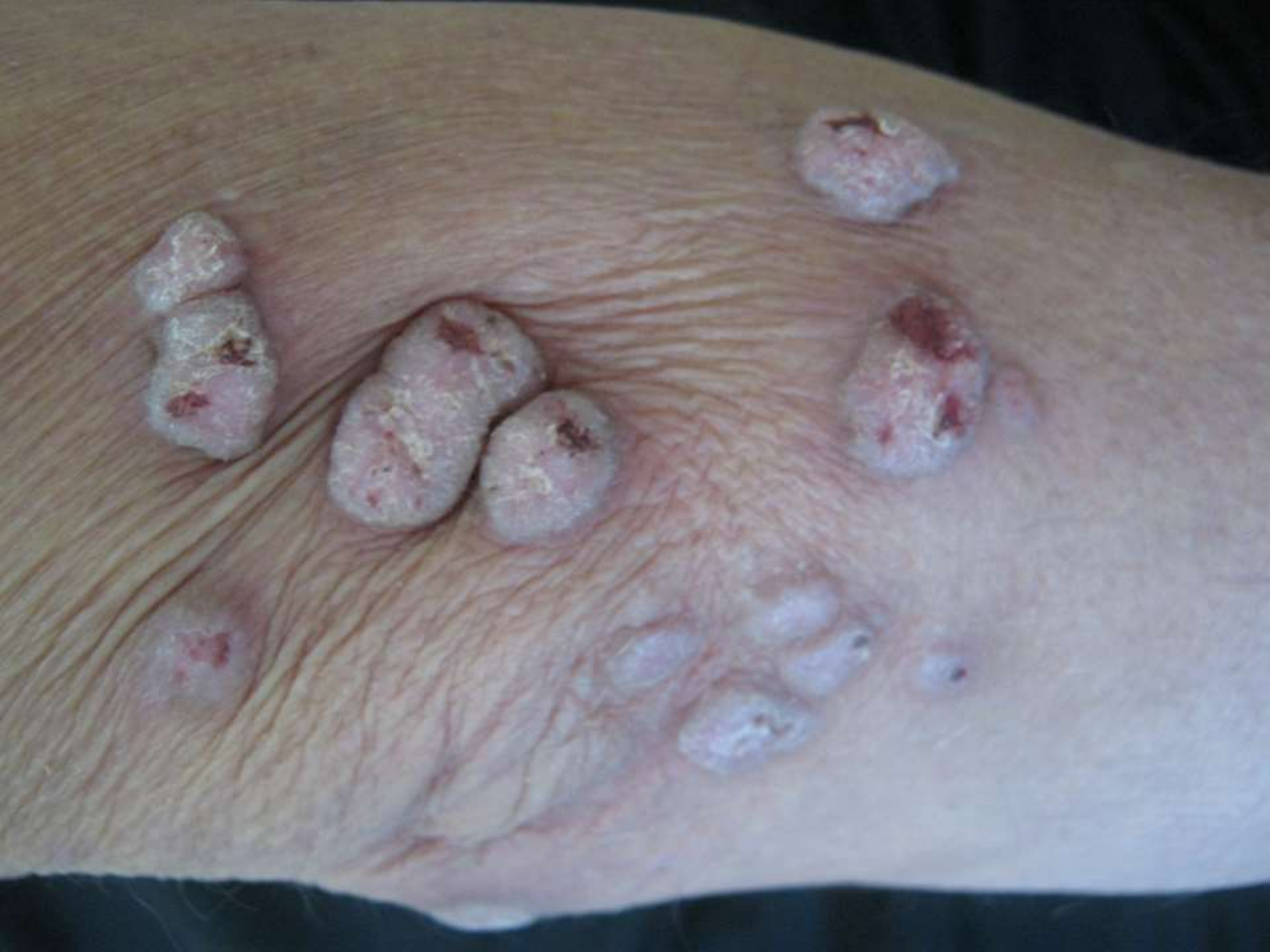
- 1. Typical form – white (pearl) papules on mucose of cheeks, tong, hard palate.
- 2. Exsudative-hyperemic – typical papules on erythematous mucose.
- 3. Erosive-ulcerative – superficial defect of papules in oral cavity. Erosion localized on central part of papule, peripheral part of element must be present. This form is torpid to treatment.
- 4. Blister form – subepidermal blisters on surface of papules of LRP.
- 5. Atypical form – papules on infiltrated mucosa, with involvement of saliva glands. желез.
- On tong may be hyperkeratosis form as leukoplakia and erosive-ulcerative form.
- LRP includes 2 syndromes:
- 1.Grinspani syndrome (erosive-ulcerative form of mucosa with diabetes and hypertension).
- 2. Little-Lassuer syndrome – follicular papules in big folds and cicatricial alopecia scalp.

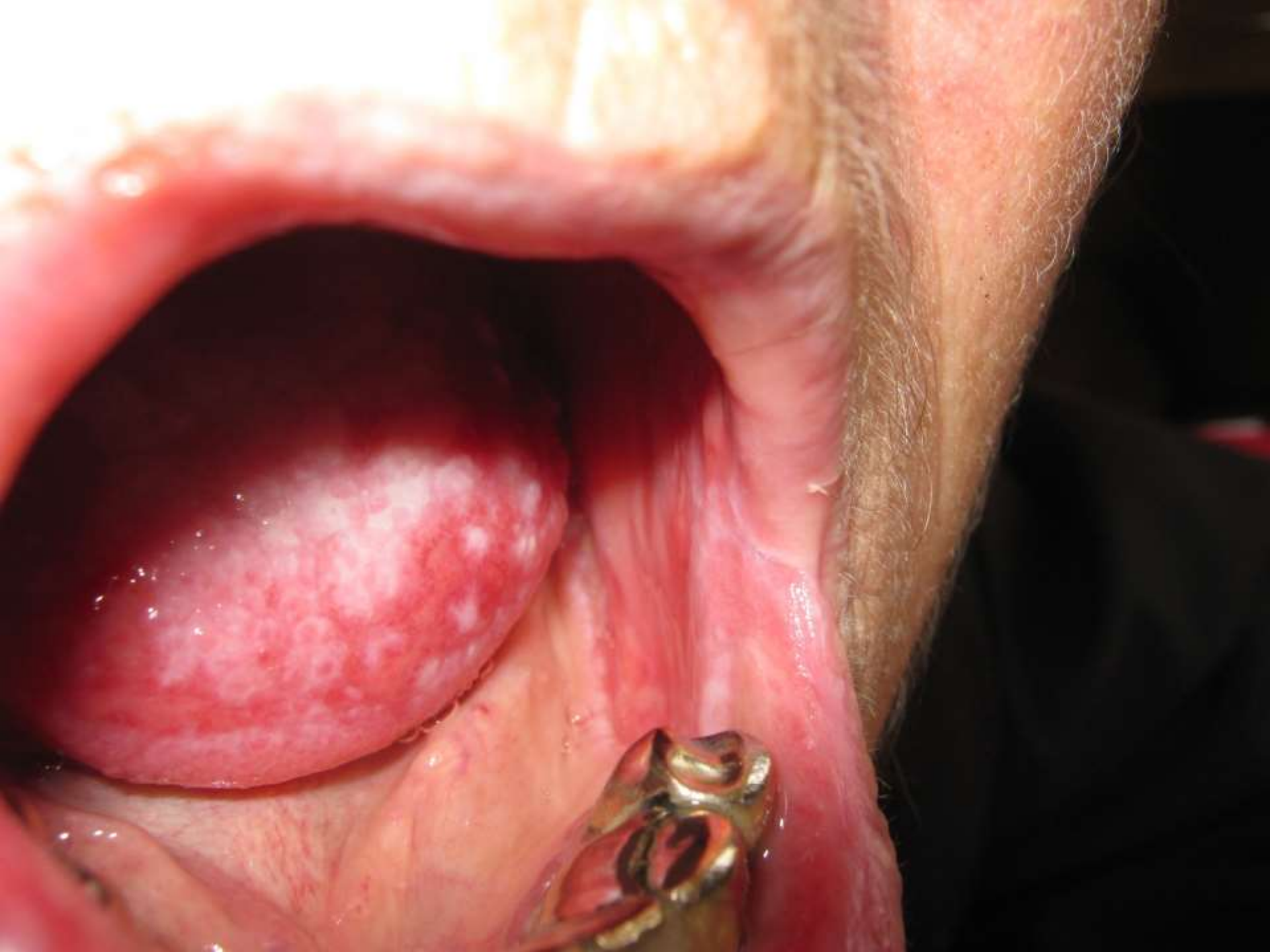
TREATMENT OF LRP

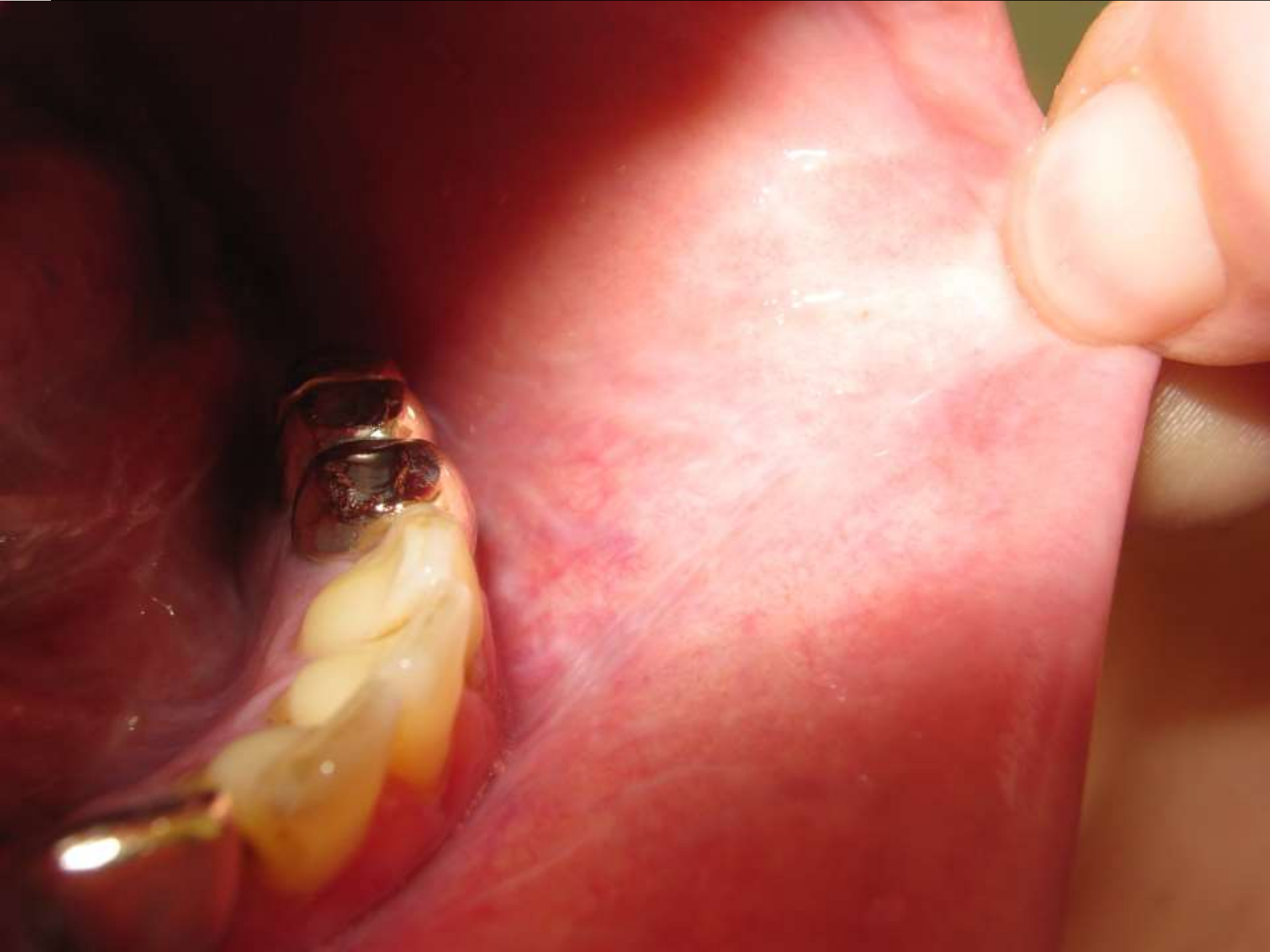
- Etiological: cure of CNS, somatic diseases, topic infections.
- Pathogenic:
- 1)systemic and topic corticosteroids,
- 2)vitamins B group and A,
- 3)antihistamins,
- 4)infusion therapy,
- 5)physiotreatment: laser treatment, baths, phonoforesis, leech therapy.











PSORIASIS

- Psoriasis is chronic multifactorial disease with genetic predisposition and dependence on external and internal factors (infections, drugs, psychological stresses, metabolic disturbances, climate, seasons.
- Clinical picture: primary element is papule of pink with silver scaling on surface, secondary – spots depigmented or hyperpigmented.
- Histological signs: acantosis, parakeratosis, papillomatosis.

CLASSIFICATION

- There are two classifications of psoriasis:
- according to stage and according to clinical form.
- 1)progressive stage (appearance of new papules, enlargement of others, Kebners phenomenon;
- 2)stationary stage (absence of of new papules, enlargement of others, Kebners phenomenon;
- 3)regressive stage (disappearance of papules, secondary macules).

CLINICAL FORMS

- 1)psoriasis vulgaris (typical papules on skin of scalp, trunk, extensors of limbs);
- 2)psoriasis exudative (on surface of papules crusts and scales, location in big folds, itch;
- 3)psoriasis pustulosa (Zumbusch of all skin, Barber of palms and soles);
- 4)erythrodermia (universal redness of skin, severe scaling);
- 5)psoriatic arthritis;
- 6)psoriasis of nails.

TREATMENT

- Etiological: removal of factors, witch participate in pathogenesis of psoriasis.
- Pathogenic: cytostatic treatment, antiiflamative treatment.
- Symptomatic antiitch treatment, analgetics in arthralgia.
- Prognosis is individual and depends on many factors.