RHEUMATIC DISEASES IN ELDERLY PATIENTS

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How do we define an elderly patient?

- An elderly patient is at least 65 years old (65+).
- Its heterogeneity results from the individual course of the aging process and from the occurrence of other chronic diseases (leading to organ dysfunction) and the use of other drugs in each person.
- Psychosocial factor plays an increasingly important role - it is important for the occurrence and course of diseases (belonging to a group, self-interest, sense of usefulness for society) - e.g. the feeling of loneliness in an elderly person despite the lack of real indicators of loneliness is conducive to the occurrence of depression.
- In practice, therefore, among the elderly there are:
  - healthy people, often leading an active lifestyle (mainly people in the age group 65–74);
  - fit patients despite the presence of numerous chronic diseases;
  - patients with various degrees of functional disability.

Different classifications

WHO classification:

- 60-75 years of age
- 75-90 years of age
- > 90 years of age

In the United States:

- "Younger Elderly" - 65-75 years of age
- "Older Elderly" > 75 years of age

Rheumatic polymyalgia - symptoms

- A disease syndrome of unknown etiology occurring in people > 50 years of age.
- Symptoms: pain in the muscles of the shoulder girdle, pelvic girdle and neck, sometimes increasing at night; morning stiffness lasting ≥30 min;
- The pain may be unilateral at first, then it covers symmetrical areas, sometimes it prevents or makes it difficult to lift the upper limbs;
- Often accompanied by arthritis - especially in the knee, sternoclavicular and hip joints. Pasty oedema of the hands and feet may appear; muscle weakness, later they may atrophy and develop into contractures;
- General symptoms - low-grade fever, weight loss, depression.
- Giant cell arteritis coexists in ~ 20% of patients. In most cases symptoms resolve after treatment, and relapses are rare.

Rheumatic polymyalgia - symptoms

Rheumatic polymyalgia – auxillary examinations

- Blood test: accelerated ESR (usually> 100 mm after 1 h, only extremely normal or slightly accelerated), increased CRP and plasma fibrinogen concentration - better correlation with the severity of clinical symptoms than ESR and CRP), moderate anemia normo- or hypochromic, thrombocythemia, eosinophilia, slight elevation of liver enzymes, especially serum alkaline phosphatase.
- Ultrasound reveals synovitis of the affected joints, bursae and tendon sheaths.

Rheumatic polymyalgia – treatment

- GCS: p.o. prednisone 12.5–25 mg / d (or other GCS in an equivalent dose) should lead to clinical improvement within 2-4 weeks, often within a few days (pain and stiffness disappear, ESR and CRP normalize later).
- In patients with symptoms of giant cell arteritis, immediately use prednisone at a dose of 1 mg/kg/day.
- In the case of an increased risk of side effects of GCS (comorbidities, simultaneous use of NSAIDs) consider early addition of GCS to methotrexate at a dose of 7.5–10 mg / week. (with folic acid 5-15 mg / week).
- NSAIDs may be useful after the end of GCS treatment in the case of persistent minor muscle or joint problems.

Case study #1

- A 67-year-old patient admitted to the Rheumatology Department with pain in the hip girdle and shoulders that had been going on for 2 months. Chronic diseases: COPD (well controlled).
- In the laboratory tests performed, ESR 85 mm/h, CRP 42 mg/l. RF, anti-CCP (-) and uric acid - normal.
- Chest X-ray - emphysema and after specific calcifications in the apex of the lungs. The abdominal ultrasound - an enlarged prostate gland to 38 ml (PSA normal).
- Methylprednisolone at a dose of 12 mg/day was started on suspicion of rheumatic polymyalgia. During the follow-up visit after one month, the patient felt much better, muscle pain was significantly less, ESR 48. A gradual reduction in the dose of methylprednisolone was planned, calcium supplementation 1000 mg/day and vitamin D3 800 units/day were ordered.

Case study #1

- After 6 months (methyloprednisolone 4 mg/day) -> an exacerbation in the form of an increase in pain, especially in the arms, ESR 45. The dose of methylprednisolone was increased to the previously used one: 6 mg and 4 mg every second day. A month later, shoulder pain was very slight, ESR 37.
- After another 5 months, another exacerbation: pain in the muscles of the shoulder girdle, elbow joints, wrists, knee joints and feet, and swelling of the hands and feet. Morning stiffness over 2 hours. On physical examination - clear compressive tenderness of both wrists, metacarpophalangeal joints and feet.
- Methotrexate at a dose of 12.5 mg / week with folic acid 15 mg / week was included in the treatment. After 15 months of treatment, the patient felt well, no exacerbations were noted. Good treatment tolerance. Inflammatory parameters low. Doses were reduced and glucocorticosteroids were eventually discontinued. The patient remains treated with methotrexate 10 mg/week. Calcium 1000 mg/day and vitamin D3 800 U/day are still supplemented.

Giant cell arteritis

- Inflammation of the arteries, often granulomatous, occurs mainly in the elderly.
- Involvement of the aorta and/or its main branches is characteristic, especially the branches of the carotid and vertebral arteries.
- The term "temporal arteritis" should not be used because this inflammation is only one (although common) manifestation of the disease and, in rare cases, may also occur in other types of vasculitis.

Giant cell arteritis - symptoms

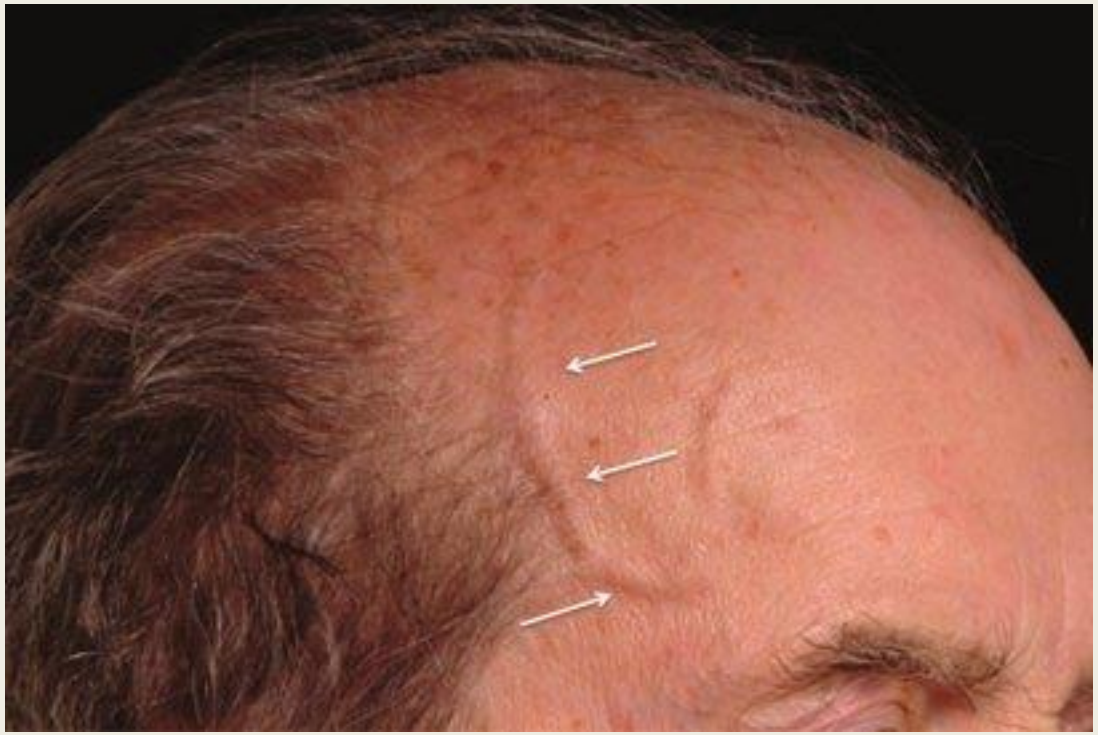
Symptoms:

- severe headache (2/3 patients), new to the patient and with scalp hyperaesthesia, occurring most often bilaterally in the temporal area or generalized, not completely resolving under the influence of analgesics, which may be accompanied by swelling and pain in the course of the temporal artery;
- in most patients low-grade fever, night sweats, weakness, anorexia, weight loss.

Giant cell arteritis - symptoms

- Ocular symptoms (in 30% of patients) - blindness (vision loss usually starts in one eye; without treatment - risk of vision in the other eye is 50% within 2 weeks (the risk of blindness is indicated by double vision and temporary lack of vision), peripheral field limitation vision; due to the risk of blindness any suspicion of disease should be considered a sudden emergency;
- Claudication of mandibule, occasional claudication and tongue ulceration;
- Neurological symptoms (up to 30%) - transient ischemia of the brain or strokes of the cerebellum or the brain, more rarely, multiple mononeuropathies; for inflammation of the aorta and its main branches, symptoms similar to those of Takayasu's disease.

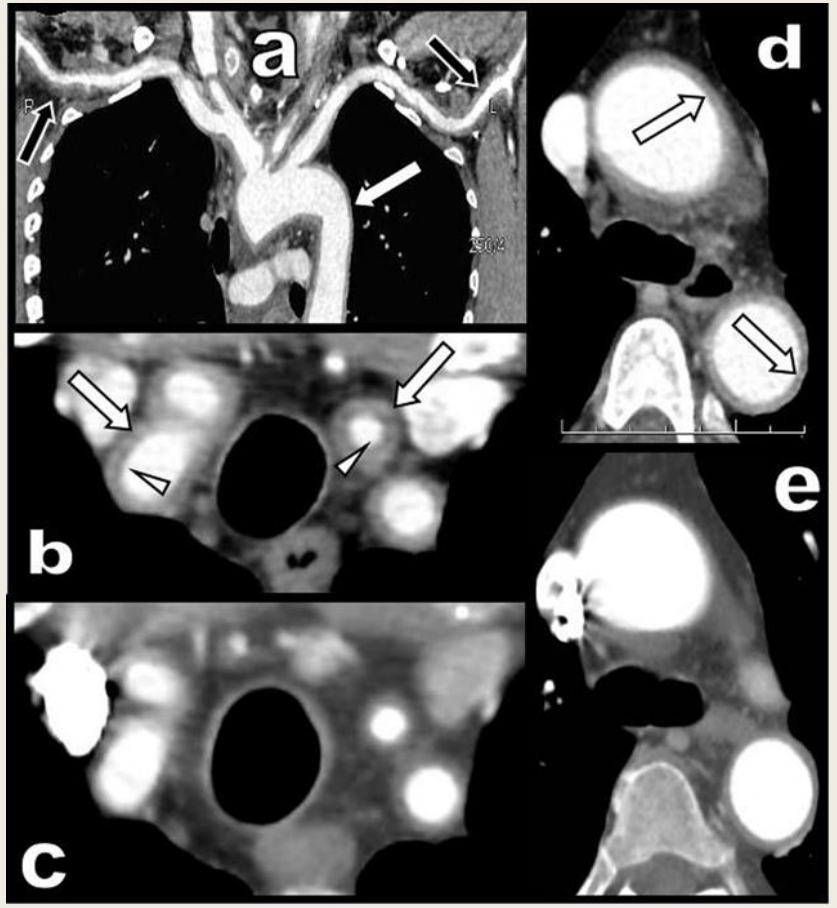
Giant cell arteritis - symptoms



Giant cell arteritis – auxillary examinations

- Blood test: increased concentration of CRP, fibrinogen, accelerated ESR (usually> 100 mm after 1 h; normal ESR [very rare] does not exclude the diagnosis), chronic disease type anemia, reactive thrombocythemia, slight increase in liver enzymes activity, especially alkaline phosphatase (~ 30%).
- Imaging tests: useful for confirming arteritis; depending on the location - ultrasound with Doppler examination (fast diagnostic path).

Giant cell arteritis – auxillary examinations



Giant cell arteritis - treatment

- Treat patients aged ≥ 50 years with acute or subacute signs and symptoms suggestive of giant cell arteritis and increased levels of CRP, fibrinogen or acceleration of ESR as an emergency and urgently refer to a specialist or specialist center for further diagnostic and therapeutic procedures.
- GCS: treatment of choice - prednisone p.o. 40–60 mg/day or other GCS in an equivalent dose, until symptoms disappear and ESR normalizes (usually within 4 weeks). In the presence of ocular symptoms at the onset of the disease, IV methylprednisolone should be used (250–1000 mg for 1–3 consecutive days).

Giant cell arteritis - treatment

- If symptoms recur, return to the last effective dose, unless ocular symptoms occur - in which case it is recommended to return to prednisone 60 mg/d or pulses of methylprednisolone i.v. (the advantage over GCS p.o. is not documented).
- In patients at particular risk of developing complications of corticotherapy (e.g. patients with diabetes or osteoporosis), consider adding tocilizumab or methotrexate (25 mg/week) to reduce the dose of GCS.

Case study #2

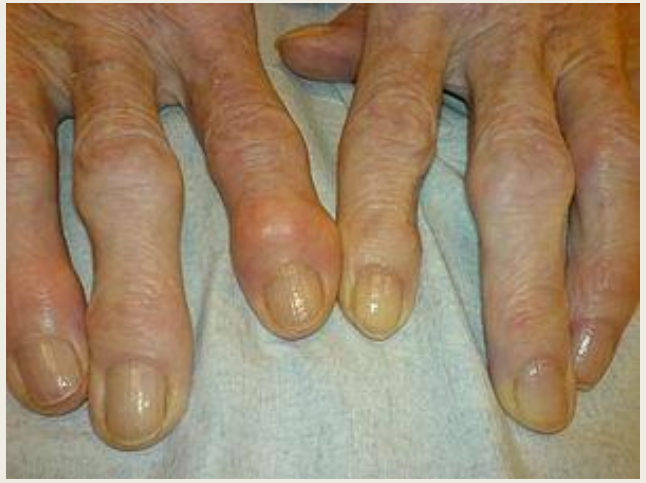
- A 67-year-old patient was admitted to the Rheumatology Department from the Emergency Department due to suspected giant cell arteritis.
- Medical history: headache in the temporal area, with a noticeable thickening of the temporal artery, swelling and pain, additionally mandibular claudication, fever - about 38.4 degrees C, pain, stiffness in the muscles of the neck and shoulder girdle. The patient denied the visual impairment.
- On admission: pupils round, even, correctly responding to light, the examination showed painfulness, swelling, thickening of the temporal arteries on both side. The abdomen was painless, without pathological resistance; no peripheral oedema. Small joints of the hands – osteoarthritis, no oedema.
- Laboratory tests showed significantly increased inflammatory parameters - CRP 208-216 mg/L, ESR 65 mm/h, slight leukocytosis, RF, ANA, ANCA, ANCA - absent.

Case study #2

- In CT of the head without signs fresh bleeding. Chest X-ray showed no abnormalities. Ultrasound - temporal arteries had thickened walls and impaired blood flow. The patient was consulted by ophthalmologist – blindness was excluded.
- Analyzing the entire clinical picture, giant cell aiteritis of the temporal arteries was diagnosed, treated with methylprednisolone iv (2.5 g in total), resulting in improvement of the general condition, reduction of pain and a significant reduction in inflammatory parameters.

Osteoarthritis

- A disease resulting from the action of biological and mechanical factors that destabilize the interrelated processes of degradation and formation of articular cartilage and the subcartilage layer of bone and ultimately involve all joint tissues. It is mainly characterized by joint pain, limited mobility in the joint, crackles and secondary inflammatory changes (e.g. joint exudate) of varying intensity, without systemic symptoms.
- Forms: primary (more frequent, cause unknown) and secondary (caused by local structural damage and abnormalities in the structure [valgus, varus] of the joint or systemic diseases).



Osteoarthritis - symptoms

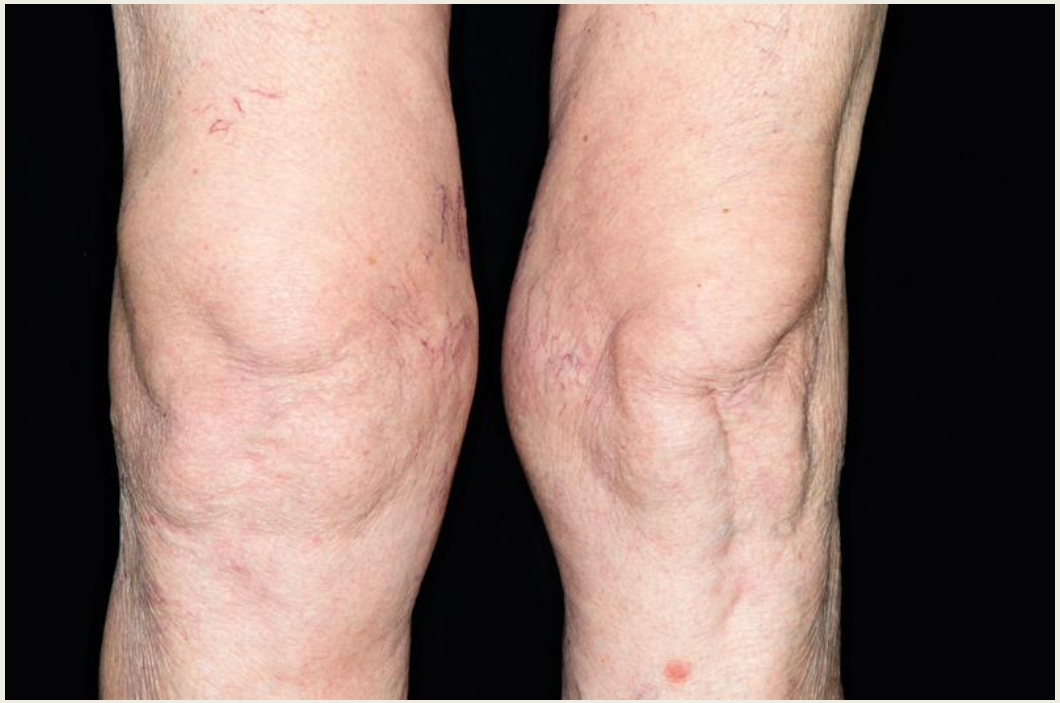
The clinical picture is usually dominated by one of the types of pathological changes (most often destruction or inadequate bone production), less often the joint inflammation. The symptoms are:

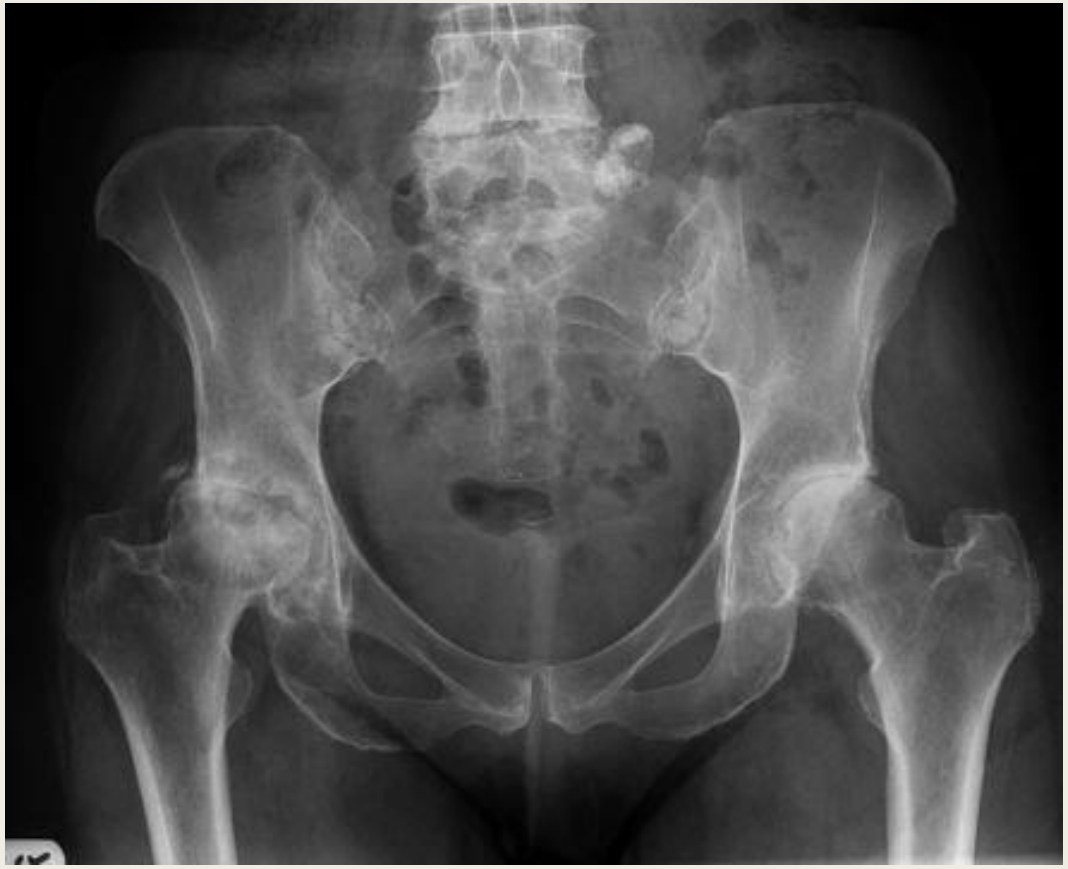
- joint pain - a dominant symptom, it usually occurs during movement in the affected joint, in the case of very advanced lesions it is severe and also occurs at rest and at night; the most characteristic feature is the greatest intensity of pain during the first movements after a period of immobility (the so-called starting pain) and a gradual reduction during subsequent movements; nocturnal pain may suggest bone marrow involvement, while pain during movement often comes from the periarticular soft tissues
- restriction of mobility in the joint, with secondary atrophy of the surrounding muscles
- rarer symptoms - dilatation and distortion of the joint contours, pain during joint palpation, crackling during movements, exudate.

Osteoarthritis – auxillary examinations

- X-ray - typical changes are: narrowing of the joint cavity caused by the destruction of cartilage, degenerative cysts (geodes) in the epiphyses due to the destruction of bone tissue, thickening of subchondral bone tissue (sclerotization), osteophytes (bone growths) at the cartilage-bone border.
- Other imaging tests (e.g. ultrasound) may be useful in differentiating from other joint and bone diseases.

Osteoarthritis – auxillary examinations





Osteoarthritis – treatment

Non-pharmacological treatment:

- Patient education.
- Diet to reduce body weight in obese or overweight patients.
- Adequate physical activity (e.g. balance exercises, tai chi, yoga).
- Kinesiotherapy to maintain the range of joint motion and muscle strength; the intensity of the pain may also decrease.
- Orthopedic equipment, e.g. a cane, crutches, limb axis correctors, knee braces (including elastic bands), external kneecap medialization, orthosis securing the thumb metacarpophalangeal joint.

Osteoarthritis – treatment

Analgetics improve quality of life and limb function, but do not significantly affect the course of the disease:

- non-steroidal anti-inflammatory drugs (NSAIDs) topically or p.o. for a limited time;
- paracetamol, tramadol;
- SNRI - duloxetine;
- intra-articular glucocorticosteroids;
- among the so-called slow-acting symptomatic drugs for OA (SYSADOA) chondroitin sulphate (for osteoarthritis of the hands);
- hialuronic acid - divergent efficacy data - ACR / EULAR not recommended, OARSI recommends application to the knee joints;
- presumably beneficial effects - hawthorn extract, ginger, incense gum, New Zealand mussel lipid complex, powdered hawthorn fruit, curcumin.

Rheumatoid arthritis

- Chronic systemic connective tissue disease of unknown origin and unknown etiology, characterized by nonspecific symmetrical arthritis, extra-articular lesions, and systemic symptoms, leading to disability and premature death.
- Depending on the presence or absence of autoantibodies in the serum (IgM rheumatoid factor and/or anti-citrullinated peptide [ACPA] antibodies), serologically positive or negative form of disease is distinguished.

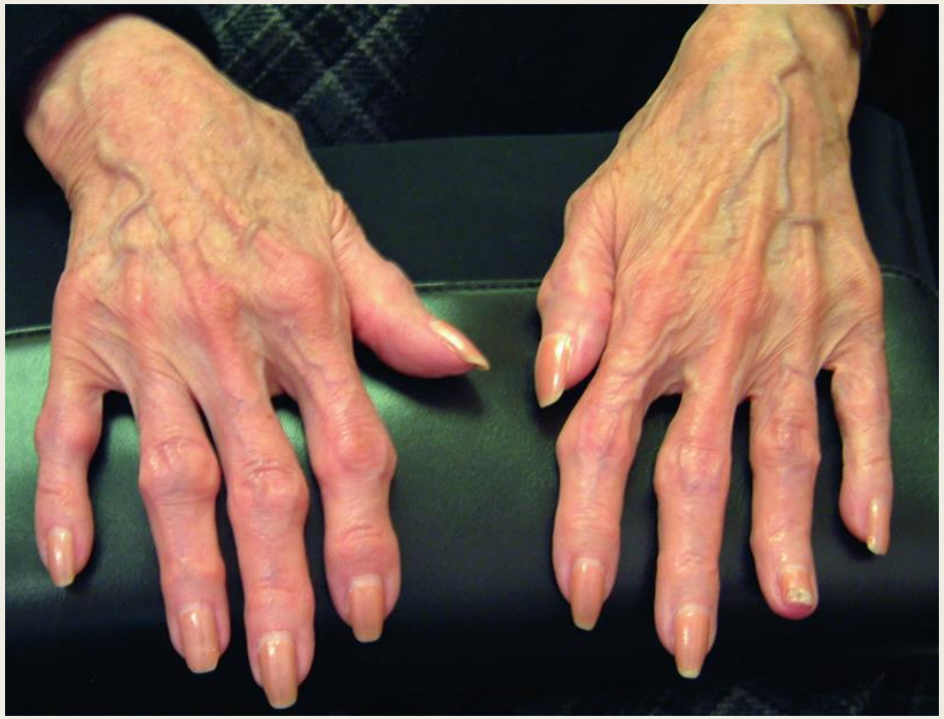
Rheumatoid arthritis

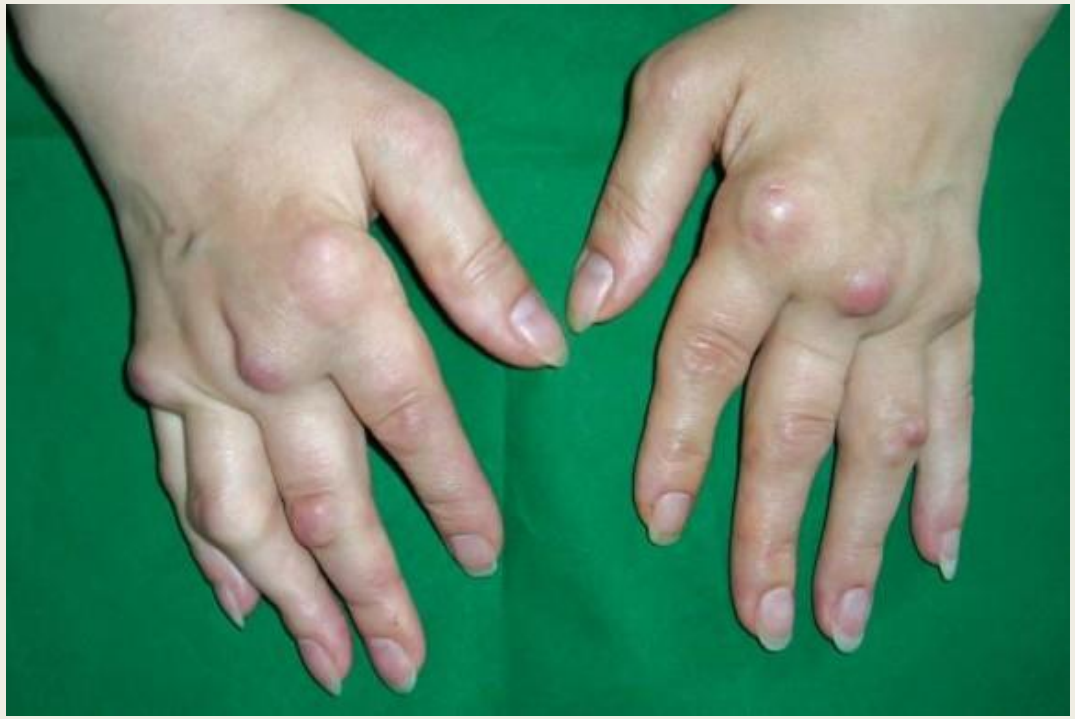
- Women are affected 3 times more often than men. Peak incidence in the 4th and 5th decade of life; possible late onset RA.
- In ~ 70% of patients periods of exacerbation and relative remissions occur with progressive joint destruction; in ~ 15% mild course with moderate disease activity, involvement of several joints and their slowly progressive destruction; in ~ 10% long-term remissions even several years; very rarely, the course of the disease is episodic (so-called palindromic) or self-limiting.
- Spontaneous remissions are more common in men and elderly patients.
- Usually the disease develops insidiously, within a few weeks; in 10–15% of patients the symptoms appear suddenly within a few days (in these cases the joint involvement may not be symmetrical).
- In> 70% of patients with active seropositive RA (IgM and/or ACPA RF serum) affecting multiple joints, significant joint damage occurs within 2 years.

Rheumatoid arthritis - symptoms

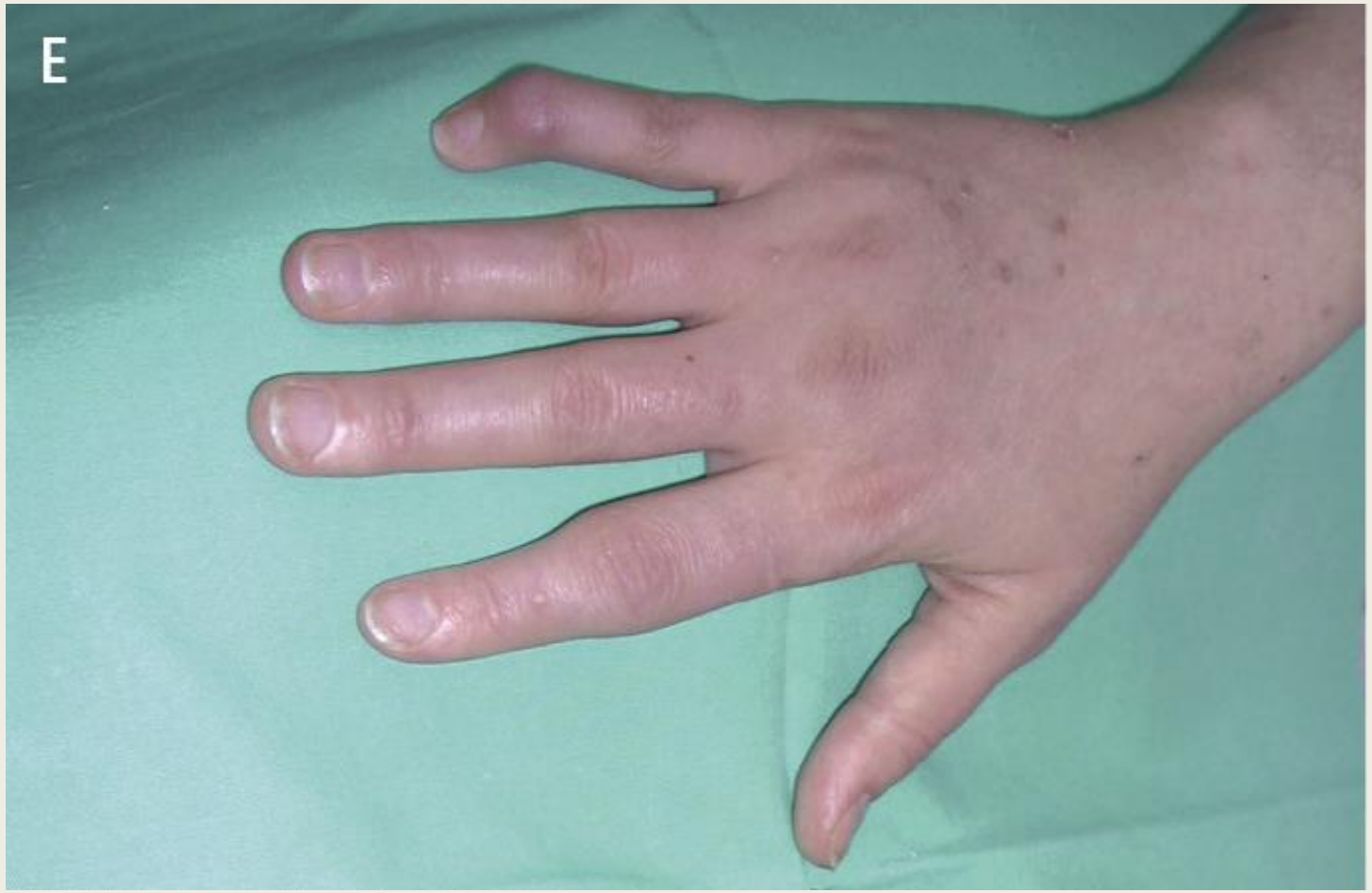
- Characteristic symptoms: symmetrical pain and swelling in the joints of the hands and feet, less often arge joints (e.g. knee or shoulder joints); morning stiffness of varying duration, usually> 1 hour.
- General symptoms: low-grade fever, muscle pain, fatigue, anorexia, weight loss.
- Movement system changes: arthritis usually symmetrical; the wrist joints, the joints of the hands and feet affected early in the course of the disease. The proximal interphalangeal, metacarpophalangeal and metatarsophalangeal joints are the most commonly involved. Subsequently, the knee, shoulder, elbow and hip joints may be affected. The joints of the upper limb (especially the wrist) are more often affected than those of the lower limb. An atypical onset is possible - inflammation of one joint or in the form of palindromic rheumatism.
- In the early stage of the disease, the following findings are noted: a slight increase in warmth (without skin reddening!). Pain during joint palpation and periarticular tissue oedema, joint effusion. May be accompanied by: tendon sheath inflammation and synovial bursitis, changes in the tendons and ligaments.

Rheumatoid arthritis - symptoms





Rhaumatoid arthritis – auxillary examinations



Rheumatoid arthritis - symptoms

Extra-articular changes: often multiorgan, mainly in the serologically positive form of RA with severe and long-lasting course:

- 1) rheumatoid nodules - subcutaneous, painless, on the extension area, mainly on the forearms, also in places exposed to pressure (e.g. buttocks), in tendons, above the joints; they also arise in internal organs;
- 2) circulatory system - pericarditis (late in the disease; exudate often clinically silent), changes in the myocardium and valves (rheumatoid nodules, cardiomyopathy), pulmonary hypertension, atherosclerosis and thromboembolic events (cardiovascular events are the most common cause of death in RA patients);
- 3) respiratory system - pleurisy (exudate, often asymptomatic), rheumatoid nodules in the lungs (they may be subject to fibrosis, calcification or infection), obliterating bronchiolitis and pulmonary fibrosis;

Rheumatoid arthritis - symptoms

- 4) eye - dry exfoliative keratoconjunctivitis in the course of secondary Sjögren's syndrome, scleritis and episcleritis
- 5) kidneys (mainly related to the side effect of the drugs used) - interstitial nephritis, pyelonephritis, secondary amyloidosis (a complication of long-term active inflammation)
- 6) other - inflammation of small and medium vessels (may lead to necrosis of the distal segments of the fingers, skin, internal organs); nervous system: carpal tunnel syndrome, polyneuropathy (mainly in the course of vasculitis), mononeuritis multiplex associated with vasculitis, pressure roots of the spinal nerves as a result of the destruction of the joints of the cervical spine, enlargement of the lymph nodes in the submandibular, cervical, axillary and elbow areas; splenomegaly (with leukopenia [neutropenia] occurs in Felty's syndrome).

Rheumatoid arthritis - symptoms

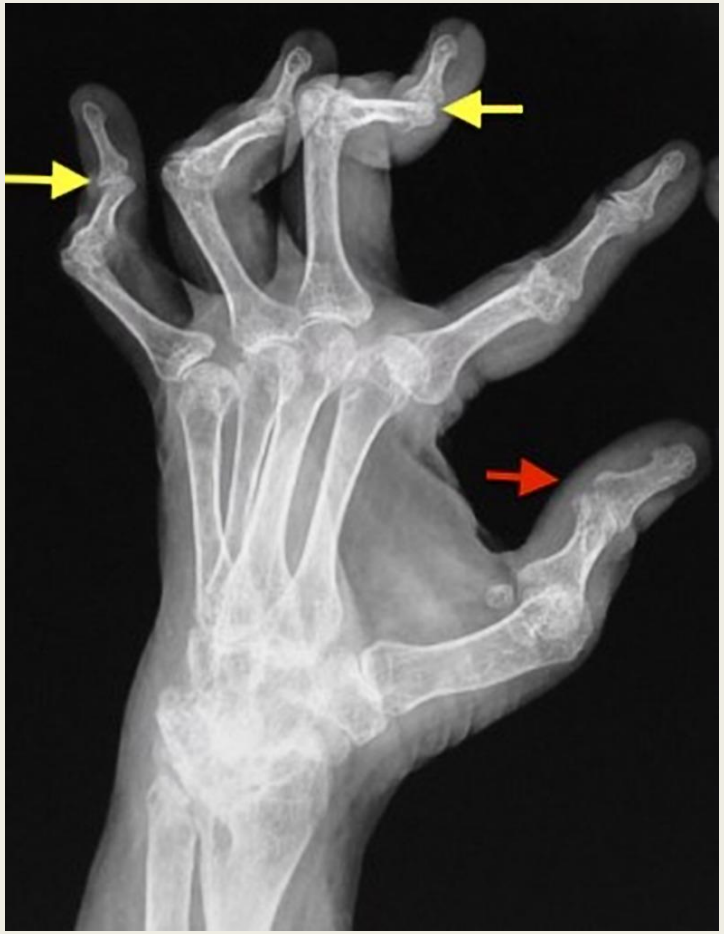
Patients with joint pain without other signs of inflammation and no other disease that may be causing the pain are at high risk of progressing to RA if ≥3 of the 7 criteria:

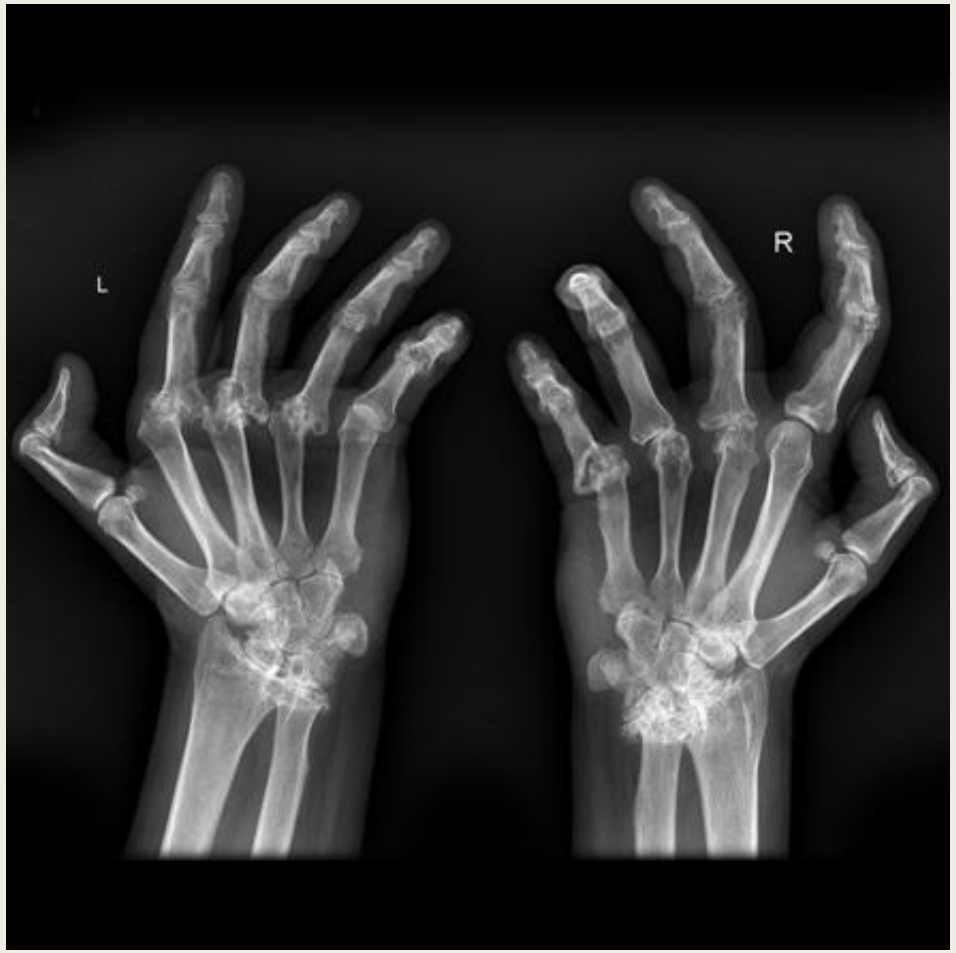
- 1) medical history
  - recent onset (<1 year) joint symptoms
  - symptoms from the metacarpophalangeal joints
  - duration of morning stiffness ≥60 min
  - greatest severity of symptoms early in the morning
  - RA in a 1st degree relative
- 2) physical examination
  - difficulties in clenching the hand into a fist
  - positive compression test of the metacarpophalangeal joints.

Rheumatoid arthritis – auxillary examinations

- Laboratory tests: ESR> 30 mm after 1 h, increased concentration of fibrinogen and CRP, normocytic and hypochromic anemia, slight leukocytosis, thrombocythemia (in a very active form of the disease) or thrombocytopenia (as a drug side effect), increased concentration of α globulin; rheumatoid factor (RF) IgM in blood in ~ 75% of patients, anti-citrullinated peptide [ACPA] antibodies.
- X-ray of the joints - X-ray abnormalities depend on the period of the disease (osteoporosis, joint cavity narrowing, geodes, erosions, nodules). Chest X-ray – interstitial lung disease, lung fibrosis.
- Ultrasound allows to detect synovitis and the presence of joint effusion in small and large joints and allows to detect erosions of the articular surfaces earlier than X-ray; in the tendons, it may reveal a loss of fibrous architecture, a cyst, or a tendon rupture.

Rhaumatoid arthritis – auxillary examinations





Rheumatoid arthritis – treatment

- Pharmacological treatment (DMARDs, GCS, NSAIDs)
- Rehabilitation
- Use in any period of the disease:
  - 1) kinesiotherapy - increasing muscle strength, improving physical fitness, preventing contractures and deformities, avoiding disability
  - 2) physical therapy - electro, laser, thermo-, cryotherapy, massages and balneotherapy have an analgesic, anti-inflammatory and muscle-relaxing effect
  - 3) psychological support.

Case study #3

- A 78-year-old patient with arterial hypertension, COPD, and osteoarthritis was admitted with suspicion rheumatoid arthritis.
- Patient complained of pain lasting for 4 months, oedema of small joints of the hands and wrist joints, pain in shoulder and knee joints, with intensification of symptoms at night/in the morning, accompanied by a feeling of morning stiffness.
- Outpatient inflammation parameters were found to be elevated and rheumatoid factor was present. Methylprednisolone was included in the treatment at the initial dose of 16 mg/day, with reduction recommended.
- Physical examination on admission revealed MCP and PIP edema of both hands.

Case study #3

- The chest X-ray showed fibrous changes in the upper field of the right lung, irregular longitudinal shadows (calcifications/post-inflammatory changes) in the lower field of the right lung, and underlined connective tissue stroma.
- In X-ray of the cervical spine and knee joints features of degenerative changes; X-ray of the hands - narrowing of the interphalangeal joint of the proximal and distal finger III, compulsive positioning in flexion, moreover, small subperiosteal cysts, degenerative changes in the left carpal-metacarpal joint I.
- The right hand ultrasound shows a trace of exudate in the MCP II, III and IV as well as in the proximal interphalangeal joints, in the tendon sheath. III finger flexors - features of chronic inflammation - a slightly overgrown synovial membrane and slight exudate, in the tendon sheath of the flexor muscles of the V finger also an overgrown synovium.

Case study #3

- In laboratory tests: ESR 17 mm/1 h, CRP 11.3 mg/L, RF present, positive CCP antibody titer, negative ANA, SS-A, SS-B, Sm, dsDNA, Borelia IgM, IgG, p -ANCA, c-ANCA, no HCV, HBV infection.
- Rheumatoid arthritis was diagnosed and sulfasalazine was proposed for further treatment. Physical rehabilitation recommended, periodic rheumatological control.
- In addition, proteinuria was found in urinalysis. Due to the possible coexistence of amyloidosis, fiberosigmoidoscopy was performed, in which a rectal biopsy was taken for histopathological examination.

Sjögren syndrome

- Chronic inflammatory autoimmune disease of unknown etiology with presence of lymphocyte infiltration and impaired function of lymphocytes, as well as inflammatory changes in many organs. There is a primary syndrome (40% of cases) and a secondary syndrome (in the course of other diseases, usually RA).
- More than 90% of patients are women. Peak incidence ~ 50 years of age.

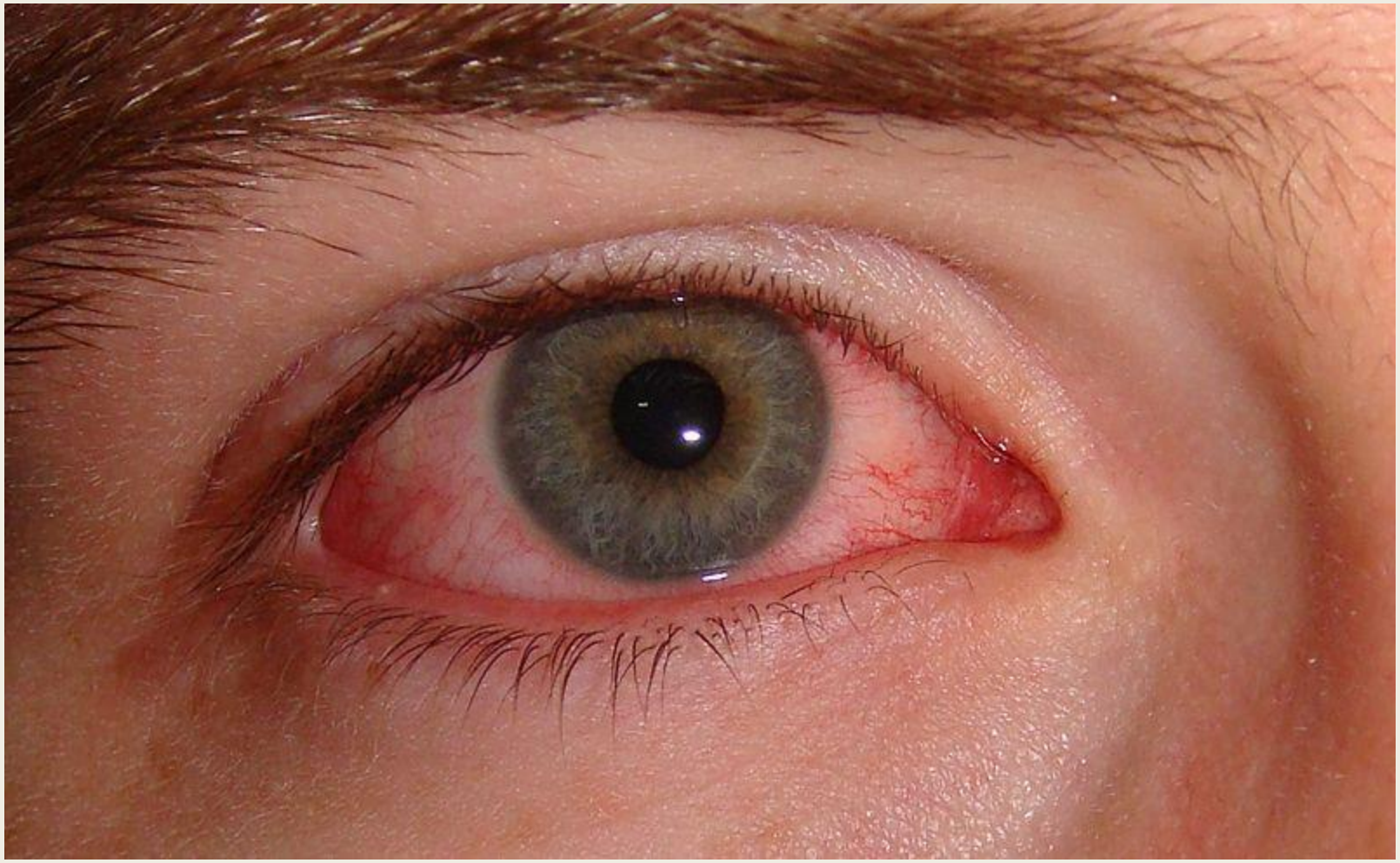
Sjögren syndrome - symptoms

- Symptoms related to changes in the glands:
- 1) lacrimal - bilateral dryness of the cornea and conjunctiva (keratoconjunctivitis sicca) felt as the presence of "sand" under the eyelids, burning, scratching; hypersensitivity to light, wind, cigarette smoke; conjunctival hyperemia;
- 2) salivary - a feeling of dry mouth, difficulty in chewing and swallowing food, speech difficulties, loss of taste, rapidly progressing tooth decay, difficulties in using dentures; enlargement of the parotid and submandibular salivary glands, inflammatory changes in the oral mucosa.

Sjögren syndrome - symptoms

- Extra-glandular symptoms: systemic symptoms such as fatigue, low-grade fever, joint and muscle pain, sometimes RA-like arthritis, mild myopathy symptoms; Raynaud's phenomenon (~ 40%); lymphadenopathy (20%); lung lesions (up to 20%; usually oligosymptomatic or asymptomatic; rarely lymphocytic pneumonia, nodular lesions or lymphoma); changes in the kidneys (up to 15%; mainly interstitial inflammation, less often tubular acidosis, sometimes urolithiasis and impaired renal function); pancreatitis, liver enlargement; primary sclerosing cholangitis; inflammation of small skin vessels in the form of purpura, urticaria, ulceration; peripheral neuropathies; dry and itchy skin (up to 55%), autoimmune thyroiditis (common, but mostly asymptomatic).

Sjögren sydrome - symptoms



Sjögren syndrome – auxillary examinations

Auxiliary research

- Blood tests: hypergammaglobulinemia (in 80%), monoclonal gammapathy (4–22%), rheumatoid factor (60%); anemia (25%), leukopenia (10%).
- Ultrasound (most useful) - allows you to assess the size and structure of the parotid and submandibular glands, detect cysts in the salivary glands, or enlargement of lymph nodes.
- Ophthalmological examinations: Schirmer's test to assess tear secretion - a strip of sterile 5 × 30 mm filter paper with a rounded edge at one end is folded and placed under the lower eyelid so that it does not touch the cornea; correctly, the length of the tissue section moistened with tears after 5 min> 5 mm.

Sjögren syndrome – treatment

Dryness syndrome

- 1) dry eye syndrome - use: "artificial tears" containing lubricants and agents increasing viscosity in a liquid or gel, at night in an ointment; soft contact lenses; Topical NSAIDs or GCs can be used ≤2–4 weeks; if artificial tears are ineffective, consider topical application of cyclosporin A drops, or serum drops, or if they are ineffective - tear duct plugs;
- 2) oral cavity - in mild dysfunction, start with non-pharmacological stimulation (sugar-free acidic candies, lozenges, xylitol, sugar-free chewing gums); in moderate dysfunction - from muscarinic receptor antagonists (e.g. pilocarpine 5 mg every 6 h), and in the case of intolerance or contraindications - from acetylcysteine or bromhexine; in severe dysfunction, treatment that stimulates the salivary glands is ineffective - saliva substitution is recommended. Recommend avoiding alcohol and smoking, and good oral hygiene;

Sjögren syndrome – treatment

- Musculoskeletal pain: Paracetamol or NSAIDs for ≤10 days;
- Systemic disease: treatment depends on the organs involved and the severity of the lesions; acute salivary gland inflammation → NSAID for several days; inflammation of> 5 joints and diffuse annular erythema → hydroxychloroquine and GCS.
- Secondary Sjögren's syndrome: treat the underlying disease.

Case study #4

- A 65-year-old patient was admitted for rheumatological diagnostics. Medical history: erythema spots on the skin, first on the lower and upper limbs, and then on the abdomen; topical and oral GCs - no improvement, cefuroxime - no improvement, clobetasol - short-term improvement. Hospitalized in the Department of Dermatology, where a biopsy of the changed skin was taken - hypergic purpura was diagnosed.
- On admission, the patient complained about eyes and mouth dryness. Good general condition, spots on the skin of lower and upper limbs and abdomen. Schirmer's test positive (<=5mm) both in left and right eye.
- Chest X-ray and abdominal ultrasound - no significant abnormalities.

Case study #4

- In laboratory tests: accelerated ESR (53mm/1h), normal CRP, positive RF factor, positive antybody titre: ANA, SS-A/Ro, SS-B/La; negative antibody titre: CCP, dsDNA, Sm, cardiolipin, beta-2-glycoprotein I, lupus anticoagulant, RNP, p-ANCA, c-ANCA; increased concentration of IgG and IgA.
- During the stay, the patient was consulted by ophthalmologist in order to qualify for the treatment, and a biopsy of the lower lip salivary gland was planned. Sjogren's syndrome was diagnosed. Introduction of hydroxychloroquine treatment was recommended after an ophthalmological consultation.

Scleroderma

- Systemic connective tissue disease characterized by progressive fibrosis of the skin and internal organs (leading to their failure), impaired blood vessel morphology and function, and abnormalities in the immune system. Etiology is unknown.
- Women are affected 3–4 times more often than men. The peak between 30 and 50 years of age.

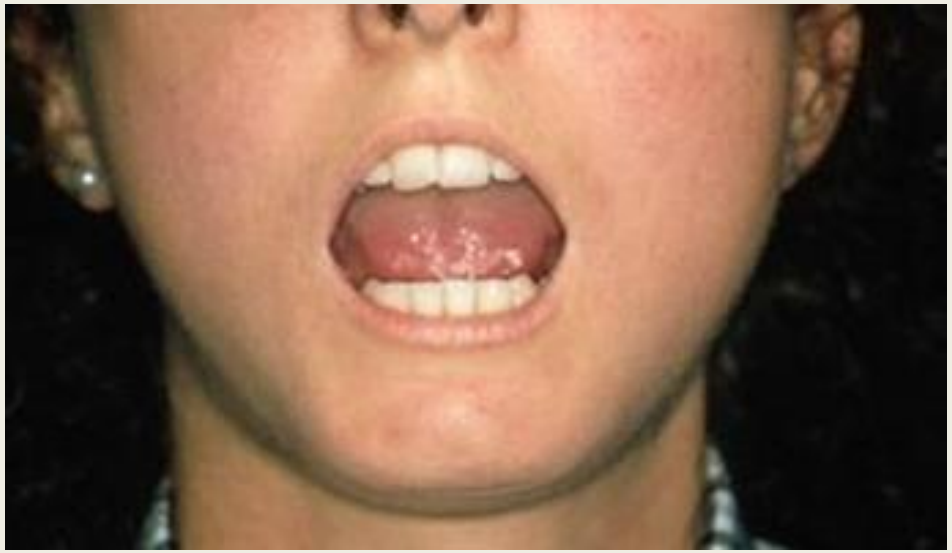
Scleroderma - symptoms

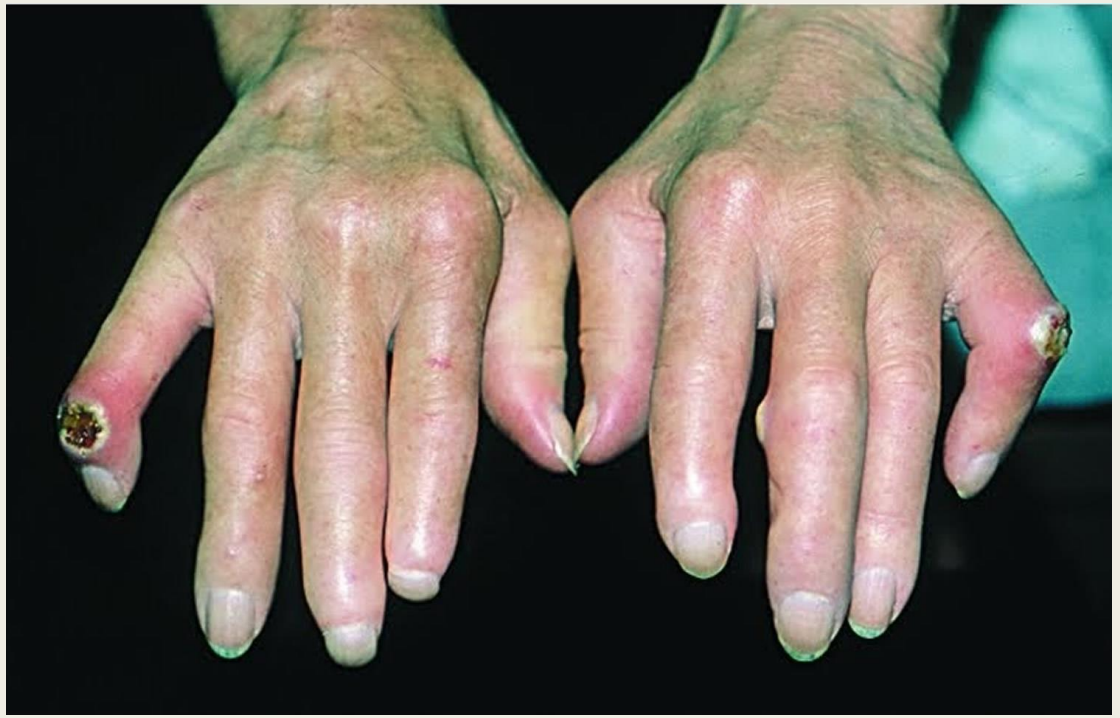
- Raynaud's phenomenon: in nearly 100% of patients with LSC and> 90% of patients with DSC.
- Skin changes: go through 3 phases - oedema, induration and atrophy; fingers of hands: initially sausage-like appearance (limited bending), then partial contracture (symptom of a tight glove); in LSC, pain associated with easily formed skin lesions and difficult to heal ulcerations, most often on the fingertips; atrophy of the fingertips, nails and shortening of the distal phalanges;
- masked face, with tight skin, slim and hooked nose, narrow mouth with radial furrow around, inability to open the mouth wide and stick out the tongue;
- skin hyperpigmentation (discoloration surrounded by an area of hypopigmentation);
- telangiectasia, especially in the skin of the face (including mucous membranes); calcifications (most often in the skin of the fingers and the extended surfaces of the elbow and knee joints; severe calcification,
- itching (mainly in dSSc).

Scleroderma - symptoms

- locomotor system: joint pain of variable location, usually symmetrical, often severe; morning stiffness, most commonly fingers, wrists, elbows, and knees; short-term swelling of the joints; restriction of mobility in the joints due to hardening of the skin; friction with movements caused by changes in the tendons (in severe forms of dSSc);
- muscle symptoms: pain and weakness;
- gastrointestinal symptoms: loss of lingual papillae, thickening of the mucosa covering the alveolar processes, gastroesophageal reflux disease, flatulence, abdominal pain, malabsorption syndrome, bleeding from vascular changes, primary biliary cirrhosis;
- respiratory symptoms: rapid breathing and shortness of breath, chronic dry cough;
- cardiovascular symptoms: arrhythmia and conduction disturbances, pulmonary arterial hypertension, left ventricular dysfunction, ischemic heart disease, pericardial disease, or myocarditis;
- kidney symptoms: scleroderma renal crisis.

Scleroderma - symptoms





Scleroderma – auxillary examinations

Auxiliary examinations:

- Blood tests - moderately accelerated or normal ESR (clearly accelerated ESR usually indicates organ complications), anemia (usually minor, increases in the case of the development of the malabsorption syndrome and progression of changes in the kidneys), hypergammaglobulinemia (increase in IgG and IgM), serum RF (in 20-30%);
- X-ray of the hands - may reveal osteolysis of distal phalanges (in the early stage, the image of sharpened pencil, then complete resorption of the distal phalanx), subluxation in interphalangeal joints, calcification; less frequently, similar changes in the X-ray of the feet.

Scleroderma – auxillary examinations

- Gastroscopy: gastroesophageal reflux and telangiectasias in the esophagus; scattered vascular changes in the stomach, mainly in the glands (single or multiple telangiectasias, imitating a watermelon stomach).
- Respiratory tests: restriction in advanced interstitial lung disease.
- ECG (disturbances of rhythm and conduction).

Scleroderma – Treatment

Treatment:

- There is no causal treatments or that are effective in inhibiting or delaying disease progression; the so-called organ specific therapy, which increases the survival rate of patients with SSc.
- In order to improve or maintain physical fitness (including contracture prevention) → physiotherapeutic procedures and kinesiotherapy (gymnastics, often preceded by paraffin compresses).
- Due to the adverse effect on skin lesions and the risk of inducing scleroderma renal crisis, do not use GCS (if it is necessary to use glucocorticoids, when life-threatening organ changes or overlap syndrome occur, monitor serum creatinine levels and blood pressure), cyclosporine, NSAIDs and drugs affecting for vascular tension such as ephedrine, ergot derivatives, β-blockers.

Case study #5

- An 82-year-old patient was admitted to the Rheumatology Department because of systemic connective tissue disease suspicion.
- Medical history: intense Raynaud's phenomenon in the hands and feet, as well as complaints of dry mouth, pain in the lumbosacral part of spine, without other symptoms of systemic connective tissue disease.
- On admission, general condition was fairly good. Oedema of the fingers, dry erosions of the fingertips of the 2nd and 4th of right hand, and the 2nd left hand, single telangiectasias. Irregular heart rate, frequency approx. 60/min.
- Laboratory tests: CRP 6.5 mg/L, ESR 11 mm/h, RF highly positive, ANA positive, the presence of Scl-70, SS-A, SS-B antibodies, decreased level of complement C3c, and increased concentration creatinine, urea.
- The echocardiography showed a high probability of pulmonary hypertension, conduction disorders.

Case study #5

- The imaging examinations showed an oval, upper-lateral shadow from the atherosclerotic aortic arch - further diagnosis required; in addition, suspicion of interstitial lung disease.
- Diagnostics was extended to CT of the chest and abdominal cavity, which revealed a 19mm x 26mm x 30mm nodular lesion in the upper lobe of the left lung, with blurred, spicular edges and a wide base adjacent to the mediastinal pleura. Surrounding vessels with infiltration features, accompanied by a pathological lymph node - for PET/bronchoscopy verification.
- Moreover, visible on both sides, mainly peripherally, basically small areas of the medullary septal thickening and frosted glass, and single cylindrical distortions of the peripheral bronchi from pulling - the image suggests interstitial fibrosis. In addition, in the lungs nodular and fibrous lesions on both sides in the apices - probably post-TBC, disseminated emphysema, a trace of fluid in the right pleura.

Case study #5

- Capillaroscopy was performed - an image uncharacteristic for the scleroderma spectrum was found. The patient was consulted by cardiologist - with no indications for the diagnosis and treatment of pulmonary hypertension. The patient did not consent to further diagnostics.
- Analyzing the entire clinical picture, systemic scleroderma was diagnosed (finger edema, finger ulceration, telangiectasia, Raynaud's phenomenon, interstitial lung disease, the presence of high-titer Scl-70 antibodies), with secondary Sjogren's syndrome. IPP included, sulodexide. The patient was referred for PET examination. The patient was discharged home at her own request.

Case study #6

- A 78-year-old patient was admitted for rheumatological diagnostics.
- RA diagnosed about 50 years ago. So far treated with: MTX, sulfasalazine, hydroxychloroquine, gold salts, GCS. Currently, MTX 20mg/week and prednisone 10mg/day.
- Patient reported increased pain in small joints of hands, feet, ankles, shoulder joints, additionally he reports the occurrence of Raynaud's phenomenon for about 2 years, periodically erosions in the oral cavity. The outpatient tests showed the presence of ANA bodies, the specification includes anti-centromeric antibodies. The right shoulder joint ultrasound revealed signs of inflammation, and MRI of the C-segment of the spine - features of multi-level discopathy with modeling of nerve structures.
- On admission: limitation of extension, abduction in the shoulder joints, palpation MCP II-V, PIP II-IV, R = L wrist joints and ankle joints, oedema of the fingers, with moderate skin induration.

Case study #6

- Laboratory tests: CRP 1.4 mg/L, ESR 5 mm/h, RF positive, anti-CCP absent, highly positive ANA body titer, presence of anti-centromeric bodies, and increased creatinine concentration, in the general urine test showed no deviation. Markers were determined: AFP, Ca19.9, Ca125, Ca15.3, CEA within the accepted standards, in electrophoresis without monoclonal protein.
- Radiologically: X-ray of the klp revealed streaked fibrosis at the base of the lung, possible discrete bilateral fibrous changes in the lower fields of both lungs, also arched widenings of the upper mediastinum on the right side, X-rays of the joints of the hands characteristic of RA. X-rays of the joints of the feet valgus of the left toe with features of subluxation; in addition, features of degenerative changes.
- Capillaroscopy showed an image that may correspond to the scleroderma pattern. The diagnostics was extended to HRCT, which showed an airy area of reticulate fibrosis.

Case study #6

- The echocardiography without signs of pulmonary hypertension, showed a slight mitral regurgitation. Using the ACR/EULAR 2013 classification criteria the rheumatoid arthritis and systemic scleroderma overlap syndrome was diagnosed (abnormalities of the nail shaft capillaries typical of scleroderma, Raynaud's phenomenon, sclerotic oedema, presence of antibodies typical of systemic scleroderma).
- Due to chronic kidney disease and the observation for malignant bladder papilloma, MTX discontinuation was recommended and the decision to start leflunomide was suspended. Betablocker was discontinued, ivabradine was implemented. It was recommended to reduce the GCS dose. The rheological preparation was included in the treatment, and it was recommended to maintain the ACEI, PPI.

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- · Źródło zdjęć: google graphics.

THANK YOU!