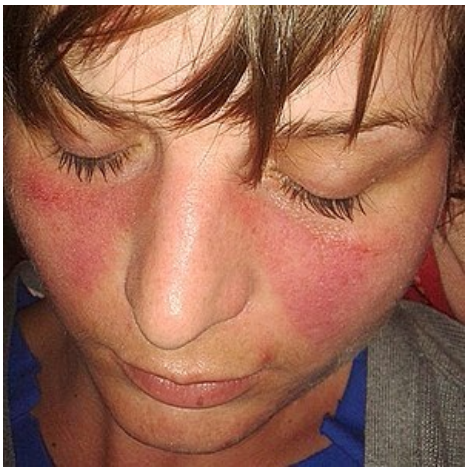
Systemic Lupus Erythematosus (SLE)

Systemic lupus erythematosus *(lupus erythematosus disseminatus)* is a serious **autoimmune disease** with a significant prevalence in women. It is characterized **by multiorgan involvement,** including organs necessary for life (especially the kidneys and brain). The basic laboratory feature is excessive overproduction of **autoantibodies to intracellular antigens**.

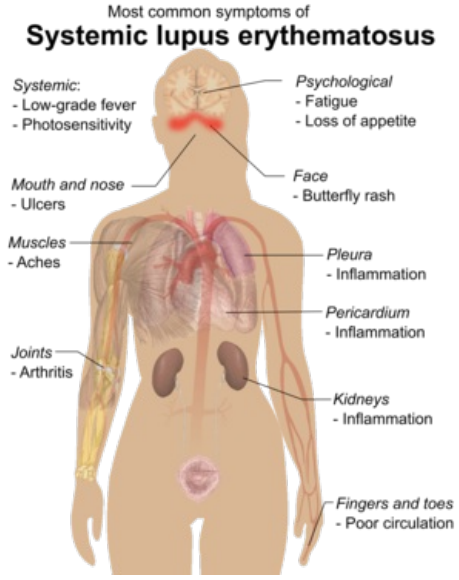


Butterfly rash in SLE patient

Etiology

- **Multifactorial etiopathogenesis**: predisposition in congenital defect of some components of complement and in HLA haplotype A1 +, B8 +, DR4 +;
- **tissue damage by autoantibodies** with the participation of a wide range of sub-mechanisms, such as cell lysis, immunodeposition, stimulation or inhibition of receptors, penetration into living cells, etc.

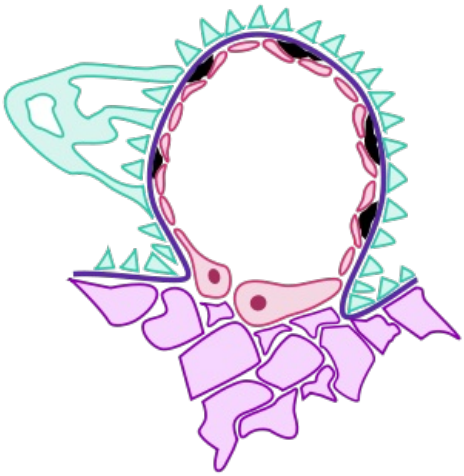
The following are particularly provocative in the manifestation of the disease: **sun exposure, drugs and estrogens** (hormonal contraceptives). The so-called drug lupus can be provoked by chlorpromazine, hydralazine, procainamide, isoniazid, penicillamine and other drugs. Hematoxylin bodies (glomeruli, endocardium and elsewhere) are a typical finding: they contain DNA from decayed nuclear material and immunoglobulin (probably antinuclear autoantibodies).



Symptoms of SLE cs

Clinical picture

- **General manifestations** (fatigue, myalgia, arthralgia, weight loss and especially temperature rise from subfebrile to intermittent sepsis-like fever);
- **arthritis and arthralgia** of the small joints of the hand (proximal interphalangeal and metacarpophalangeal joints), knees and wrists. On X-ray we find periarticular osteoporosis without erosion;
- **butterfly rash** on the face that produces a reddish, raised lesion (often itchy or painful)
- discoid lesions (sun exposure on the face or other parts of the body): these are reddish papules and areas with a less pigmented center;
- **photosensitivity** can be demonstrated in about half of patients;
- **serositis** – pericardial effusion or pleurisy;
- **lupus glomerulonephritis** is one of the most common and severe organ manifestations of the disease. It is manifested by proteinuria above 0.5 g / 24 hours, the presence of cylinders in the sediment, microscopic erythrocyturia, or pyuria (in the absence of infection). In nephrotic syndrome, proteinuria is above 5 g / 24 h with variable total edema.
  - Normal finding.
  - Mesangial lupus nephritis.
  - Focal proliferative lupus nephritis.
  - Diffuse proliferative glomerulonephritis.
  - Membranous glomerulonephritis.
  - Sclerosing glomerulonephritis.
- **acute lupus pneumonitis** – a febrile condition with cough and spotted alveolar infiltrates on an X-ray of the lungs;
- **lupus myocarditis** with pericardial effusion (serous Pericarditis), tachycardia and arrhythmias;
- **neuropsychiatric symptoms** – from depressive tuning, migraine headaches to epileptiform paroxysms, visual disturbances and psychotic states;
- **papillary swelling of the optic nerve and cotton wool deposits on the retina .**



SLE Nephritis Pathology Diagram

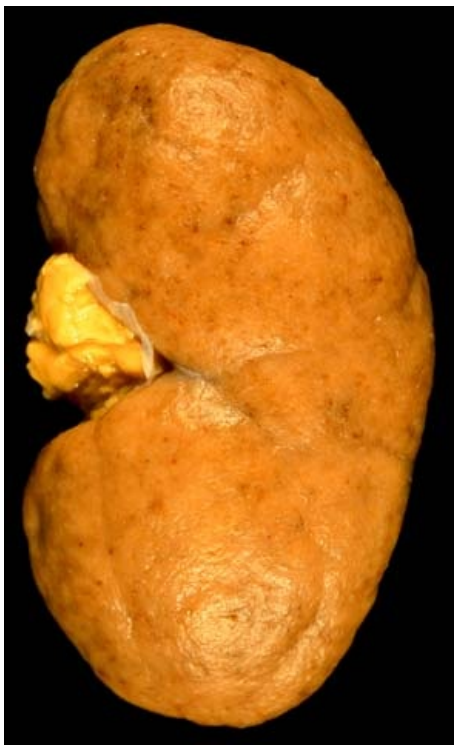
Laboratory finding

- Increased sedimentation and an increased number of acute phase proteins, mild anemia, leukopenia with lymphocytopenia, mild (rarely severe) thrombocytopenia with normal or increased megakaryocytes in the sternal puncture) is often found . Prolongation of activated partial thromboplastin time (APTT) in the current

antiphospholipid syndrome;
- detection of antinuclear autoantibodies (ANA) "overall" by immunofluorescence or enzyme-linked immunological chemistry;
- detection of double-stranded (native) DNA autoantibodies (ie anti dsDNA or anti nDNA autoantibodies).

Diagnostics

- Proof of 4 or more of the following 11 ACR classification criteria as modified in 1983:
  - butterfly rash;
  - discoid rash;
  - photosensitivity;
  - defects (ulcers) of the oral or nasopharyngeal mucosa;
  - non-erosive arthritis of two or more peripheral joints;
  - serositis,
    - pleurisy;
    - pericarditis;
  - kidney disorder;
    - persistent proteinuria above 0.5 g / 24 h;
    - cell cylinders;
    - hematuria;
  - neurological disorders;
    - convulsions (excluding other causes);
    - psychosis (excluding drug or metabolic origin);
  - hematological disorders;
    - hemolytic anemia with reticulocytosis;
    - leukopenia below $4.0 \cdot 10^9$ / l (repeated identification);
    - lymphopenia below $1.5 \cdot 10^9$ / l (repeated identification);
    - thrombocytopenia below $100 \cdot 10^9$ / l (unless drug-induced);
  - immunological disorders;
    - positive LE cells;
    - anti-DNA autoantibodies in serum at abnormal values of the assays used;
    - anti Sm autoantibodies in serum (about 10% of patients);
    - detection of aCL antibodies;
  - abnormal ANA titer by immunofluorescence test or other equivalent technique (if positive induction drugs can be ruled out.


Diffuse proliferative lupus nephritis

Therapy

- **Glucocorticoids**;
- in the active form of the disease with intestinal manifestations - pulse treatment with **methylprednisolone** (MP): it is a series of 3-5 daily or bi- daily intravenous infusions of mega doses of MP, ie usually 1 g per day. This is followed by treatment with methylprednisone or prednisone at doses that gradually decrease to a daily minimum capable of maintaining remission;
- in prognostic conditions, pulse treatment with methylprednisone is followed by pulse treatment with **cyclophosphamide** (eg lupus glomerulonephritis with a class IV biopsy finding) or after azathioprine therapy (2 mg / kg);
- in low-activity patients without obvious vital involvement, the basis of treatment is glucocorticoid po (eg prednisone in the initial dose up to 40 mg with gradual reduction) and synthetic antimalarial hydroxychloroquine 200 mg daily;
- We warn patients against sun exposure, women of childbearing age from hormonal contraception.

Lupus neonatorum

Newborns of mothers with SLE and / orSjögren's syndrome are at risk of developing neonatal lupus. It is an autoimmune disease caused by the passive transfer of antibodies from mothers to the fetus. It mainly manifests **heart and skin problems.**.

The most serious complication is a disorder of the cardiac conduction system, typically a complete AV block, which occurs in about **2% of women with anti-Ro / SSA anti-Ro/SSA** (Sjögren syndrome type A antigen) **and / or anti-La / SSB antibodies** (Sjögren syndrome type B antigen). Any degree of block may occur and may be accompanied by valve abnormalities, endocardial fibroelastosis and / or dilated cardiomyopathy. In pregnant women with anti-Ro / SSA and / or anti-La / SSB, repeated fetal echocardiography is appropriate for early diagnosis of fetal heart block and ev. initiation of dexamethasone, which crosses the placenta. Heart block may be already prenatal (usually occurring between the 18th and 20th week of pregnancy), but is often diagnosed at birth (such as bradycardia when listened to after birth). In the most severe cases, children may need a pacemaker, but most children are relatively asymptomatic in childhood and have difficulty adolescent with increased workload such as syncope.

The cutaneous manifestation of neonatal lupus is in about 4-16% of children of mothers with anti-Ro and anti-La antibodies, but sometimes with RNP (ribonucleoprotein) antibodies. Skin manifestations are usually evident at birth. Annular erythematous deposits with mild flaking are typical, appearing mainly in the hair, neck or face (typically

periorbital), but also on the trunk and limbs.

Rarely, neonatal lupus may manifest as hepatic (elevated aminotransferases and / or conjugated bilirubin, hepatosplenomegaly) or hematologic (cytopenia). Haematological disorders (haemolytic anemia, thrombocytopenia, neutropenia) may occur within the first 2 weeks of life and disappear within 2 months of life. Skin, hepatic and haematological problems are only transient and disappear spontaneously by 4-6 months of age (when maternal antibodies disappear). Rarely, neurological complications may also occur in children of mothers with anti-Ro antibodies, such as hydrocephalus, so sonographic examination of the brain is appropriate. Pneumonitis, which manifests as tachypnea or tachycardia, may also occur.

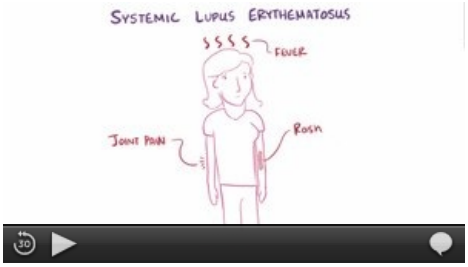
Pregnant women with SLE are at increased risk of fetal growth restriction and preterm birth.

Neonatal lupus can also occur in children of completely asymptomatic mothers who are unaware of the presence of autoantibodies, and the first manifestation may be fetal bradyarrhythmia or neonatal rash.[1][2]

SLE and breastfeeding

Most women with SLE can breast-feed, however, some medications may pass into breast milk. Immunosuppressants (azathioprine, methotrexate, cyclophosphamide, mycophenolate) are contraindicated and long-acting non-steroidal anti-inflammatory drugs are not suitable. Hydroxychloroquine passes into breast milk and increases the risk of kernikter. Breast-feeding is possible with the use of short-acting NSAIDs, antimalarials, low doses of prednisone (ie up to 20 mg / day), warfarin and heparin.[2]

Summary video



Lupus video

Links

related articles

- Henoch-Schönlein purpura
- Kawasaki disease
- Juvenile rheumatoid arthritis
- Manifestations of inflammatory rheumatic diseases on the musculoskeletal system and their surgical treatment
- Systemic lupus erythematosus / case report

Source

- ŠTEFÁNEK, Jiří. Medicína, nemoci, studium na 1.LF UK [online]. [cit. 2010-05-06]. <http://www.stefajir.cz>.
- BENEŠ, Jiří. Studijní materiály Jiřího Beneše [online]. [cit. 2010-05-06]. <http://www.jirben2.chytrak.cz>.

References

- KLENER, Pavel, et al. Vnitřní lékařství. 3. vydání. Praha : Galén, Karolinum, 2006. ISBN 80-246-1253-4.
- 1. BUYON, J P. Neonatal lupus: Epidemiology, pathogenesis, clinical manifestations, and diagnosis [online]. UpToDate, ©2019. [cit. 2020-09-20].
- 2. FEMIA, A N. Neonatal and Pediatric Lupus Erythematosus [online]. Medscape, ©2016. [cit. 2020-09-20]. <https://emedicine.medscape.com/article/1006582-overview>.

1.
2.