Lupus (Systemic Lupus Erythematosus; SLE) — Diagnosis and Treatment

The autoimmune disease lupus erythematosus belongs to the collagenoses and can show very different clinical symptoms: The symptoms range from mild conditions to severe organ manifestations which makes the diagnosis not easy sometimes. Read all the important facts about lupus erythematosus here to be prepared for the exam and the subsequent medical daily routine.

Definition of Lupus

The systemic lupus erythematosus as autoimmune disease

Lupus erythematosus belongs to the group of **autoimmune diseases** and further to the group of **collagenoses**. In the group of collagenoses, it is associated with diseases of the rheumatic spectrum disorders.
Epidemiology of Lupus

Child-bearing aged women constitute around 90% of the affected patients. Highest prevalence is observed in African-American and Afro-Caribbean women. Overall prevalence of SLE in the US varies from 20 to 150 per 100,000 women.

Etiology of Lupus

Causes of the systemic lupus erythematosus

The exact etiology is unknown but there are certain predisposing genetic factors, e.g. the surface molecules HLA-DR2 and HLA-DR3. Furthermore, external factors like hormonal changes, stress, infects, increased light exposure or drugs can “trigger” a lupus.

Pathophysiology of Lupus

The development of systemic lupus erythematosus

The pathomechanism is a precipitation of immune complexes at the basal membrane of the cell walls. This happens with connective tissue of the skin but also with vessels. The precipitations can lead to a vasculitis there and can be seen as so called “lupus band” under the microscope when staining the immune complexes with immunofluorescence stain.

These precipitating immune complexes consist of DNA, antibodies against this DNA, complement and fibrin. They develop due to a misdirected immune response against internal cell nucleus components. These are falsely distinguished as foreign by the immune system and further antibodies are produced.

In summary, genetic and environmental predisposing factors cause increased production of nucleic acids; this in turn elicits an abnormal immune response from body. The result is formation of circulating immune complexes. These immune complexes precipitate in the body tissues and cause inflammation and damage.

Clinical Picture of Lupus

Symptoms of systemic lupus erythematosus
In most cases, systemic lupus erythematosus appears with \textit{unspecific general symptoms} which can complicate the diagnosis. Fever, weakness, weight loss and swelling of lymph nodes can belong to these.

\textbf{Skin changes} that gave the disease its name (lupus= wolf, due to the wolf’s face-similar scars that stay after healing of the skin defects) manifest themselves nearly as often. The characteristic skin changes are a \textit{butterfly-like erythema} on the cheeks and nasal bridge. Vibrant, scaling papules are found at the subform of discoid lupus.

Furthermore, \textit{polyarthritis} and \textit{myositis} are some of the more common symptoms.

Eventually, SLE can lead to \textit{neurological} symptoms. In these, the peripheral nervous system is affected more commonly. The infestation becomes apparent due to a decrease of vigilance, depression, epilepsy or stroke for example.
Renal changes in systemic lupus erythematous: The lupus nephritis

The lupus nephritis that occurs in over half of the patients, is crucial for the prognosis of SLE. The complexes of DNA and anti-DNA antibodies lead to the clinical picture of a classic immune complex glomerulonephritis. This can express itself with different clinical symptoms from asymptomatic proteinuria to chronic renal failure.

It is divided into six different types by the world health organization WHO as well as International Society of Nephrology and Renal Pathology Society that are treated differently. The therapy consists of immunosuppression and an optimal blood pressure adjustment in most cases; except in case of both, minimal and extensive involvement of
Summary of the symptoms of systemic lupus erythematosus

All in all, you should think of a systemic lupus erythematosus with the following symptoms:

- Fever, weakness, weight loss
- Butterfly rash
- Polyarthritis, myositis
- Cardiopulmonary changes
- Lupus nephritis
- Neurological changes

Diagnosis of Lupus

Laboratory diagnostics for systemic lupus erythematosus

- The anti-dsDNA antibodies that are increased in 70% of the cases of lupus, and the anti-nuclear antibodies (ANA) that are nearly always increased, are characteristic. Yet, these can also be found in healthy individuals in low concentrations.
- Further antibodies can also be positive: anti-Sm, anti-Ro, antiphospholipid antibodies (APA), antihistone, antineuronal, antiribosomal etc. The circulating antibodies often lead to a decrease of the cell count of different cell types such as thrombocytopenia and lymphocytopenia.

Note:

- Increase of ANAs in 95 % of the cases
- Increase of anti-dsDNA antibodies in 70 % of the cases

Diagnostic criteria based on the American College Rheumatology

There must be at least 4 of the 11 criteria met to diagnose SLE:

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| **Dermatological** | 1. Malar rash (“butterfly rash”) with sparing of the nasolabial folds  
2. Discoid rash  
3. Photosensitivity  
4. Oral or nasopharyngeal ulcers |
| **Internal organs** | 1. Non-erosive arthritis involving at least two peripheral joints; It is usually rarely deforming.  
2. Pleuritis or pericarditis.  
4. Neurologic disorder: seizures, psychosis, and personality changes |
| **Laboratory tests** | 1. Hematologic disorders: autoimmune hemolytic anemia, thrombocytopenia, leukopenia, lymphopenia  
2. Immunological findings: anti-dsDNA, anti-Sm, or antiphospholipid antibodies  
3. Antinuclear antibodies (ANA) |
Therapy of Lupus

- There is no cure available, and complete sustained remissions are not common. Therefore, the aim is to control acute flares, suppression of symptoms to an acceptable level and prevention of organ damage.
- Patients without major organ involvement can be managed conservatively with analgesics and antimalarials. On the other hand, systemic glucocorticoids are the recommended treatment for any inflammatory life-threatening or organ-threatening manifestations of SLE.
- Cytotoxic or immunosuppressive agents, especially cyclophosphamide or mycophenolate are added to glucocorticoids to treat serious SLE. For refractory serious SLE various biologics are currently being studied. Also, studies of highly targeted interventions at various steps of immune regulation are in progress.
- The **blood pressure** has to be optimally adjusted to prevent renal damage. Furthermore, a sufficient **prophylaxis of osteoporosis** should follow.
- Other preventive strategies include providing influenza and pneumococcal vaccines and suppressing recurrent urinary tract infections.
- Controlling other co-morbidities such as dyslipidemia, hyperglycemia, and obesity is also recommended to improve SLE outcomes.

Prognosis of Lupus

Survival probability in systemic lupus erythematosus

The rate of survival is very good with optimal therapy. Lupus patients mostly die from cardiovascular complications.

Poor prognosis has been associated with high serum creatinine levels, hypertension, nephrotic syndrome, anemia, hypoalbuminemia, hypocomplementemia, antiphospholipid antibodies, male sex, ethnicity (African American, Hispanic with mestizo heritage), and low socioeconomic status.

References


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