EduVision: Journal of Innovations in Pedagogy and Educational Advancements
Volume 01, Issue 06, June, 2025
brightmindpublishing.com
ISSN (E): 3061-6972

CLINICAL MANIFESTATIONS OF THE DISEASE IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Abstract

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease characterized by multiple organ and system involvement. The clinical picture of SLE is polymorphic, which complicates timely diagnostics. The article discusses the most typical clinical manifestations of SLE with an emphasis on the systemic nature of the lesion and possible diagnostic difficulties. The immune system is a unique system of the human body that is able to distinguish "self" from "foreign", thereby providing protection against infections. Autoimmune diseases are a consequence of the activation of autoreactive T- and B-cells for reasons that are still unclear. Systemic lupus erythematosus (SLE) is a prototype of autoimmune diseases in which various systems and organs are affected. Most autoimmune diseases, including SLE, are considered as complex diseases, the manifestation of which involves environmental and genetic factors. Therapy for such diseases is not selective and extremely aggressive, which does not have the best effect on the quality of life of patients. The study of the genetic basis of autoimmune diseases is necessary to understand the pathogenetic mechanisms and discover new ways of therapy at a more advanced level - the genome level. Systemic lupus erythematosus (SLE) is a multisystem disease of unknown etiology, in which the immune system attacks its own cells and tissues. The disease is more common in women aged 15-45 years. Diagnosis is complicated by the variety of clinical manifestations and the possible involvement of almost any organ. SLE is inherited contrary to simple Mendelian laws and has a polygenic inheritance model. In some cases, SLE is associated with rare but highly specific mutations. For example, with homozygous deficiency of early components of the complement system Clq , C2 or C4. It is known that with a complete deficiency of the complement component C4, systemic lupus erythematosus develops in more than 75% of cases. However, studies devoted to the study of the genetic basis of deficiency of early complement components, including C4, are not numerous. This may be due to the labor-

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intensive nature of research into the unique region of the genome in which the C4 genes are located.

Introduction

The aim of the study. To study the role of "null" alleles of the complement component C4 gene in the pathogenesis of systemic lupus erythematosus in children of the Russian population.

Objectives of the study: To study the frequency of occurrence of "null" alleles of the C4A and C4B genes in children with SLE and in control groups in the Russian population. To determine whether the presence of "null" alleles of the C4 genes predisposes to the development of systemic lupus erythematosus. To assess the concentration of C4 protein in the blood serum of patients with C4 gene defects.

Results of the study. Systemic lupus erythematosus (SLE) is a systemic autoimmune disease of unknown etiology, characterized by hyperproduction of organ-nonspecific autoantibodies to various components of the cell nucleus with the development of immune-inflammatory damage to tissues and internal organs. The pathogenesis of SLE has not been fully studied, clinical manifestations are extremely variable, and the course is unpredictable. It is known that the disease belongs to the group of autoimmune diseases, characterized by multiorgan involvement. The disease debuts in childhood only in 10-20% of cases, but according to the results of numerous studies, SLE that debuts in childhood tends to be more severe: kidney and central nervous system damage is more common in children, organ damage occurs faster. In recent years, it has become possible to purposefully influence the genome of an organism, experimental models of knockout and transgenic animals have been created, it has become possible to selectively evaluate the function of a particular gene and the protein it encodes. Cellular and humoral links of the immune system have been studied in experimental models, which has changed the pathogenetic basis of autoimmune processes. It has been proven that in SLE, there is an interaction between the components of the adaptive immune system and the cells and tissues of the body, caused by dysregulation autoreactive B cells, which is based on the violation of immunological tolerance. The main basic drugs in the treatment of SLE are immunosuppressants. The metabolism of immunosuppressants is associated with

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the polymorphism of the genes of the P450 CYP3 family. The issue of studying the relationship between the metabolic disorder of this drug in patients with systemic lupus erythematosus in children with mutations of the genes of the P450 CYP3 family remains unstudied. All this determines the significance of the problem, the importance of conducting in-depth research in this area, the solution of which will contribute to understanding the fundamental nature of the immunological and molecular genetic mechanisms of pathogenesis, the development of preventive measures on this basis, prognosis, as well as optimization and improvement of the effectiveness of treatment of this category of patients. However, much of the information on this issue is contradictory and incomplete. Considering that in most cases of autoimmune hemolytic anemia (AIA) in SLE there is no clinical laboratory hemolysis symptom complex , diagnosis of this condition is often difficult. It is noted that hemolysis in SLE can occur in these patients with the development of secondary APS [10.8% - 46.9%]. There is virtually no information on the existence of (YDL) in SLE, although this type of AS can occur in these patients in the same way as in the population as a whole.

Anemic syndrome is a frequent manifestation of SLE and is observed in more than two thirds of patients - 76.5% . In the vast majority, the anemic syndrome is complex - a combination of autoimmune hemolytic anemia and anemia associated with iron metabolism disorder (70.8%). Only autoimmune hemolytic anemia and anemia of chronic disease are less common (9.2% of all patients with anemic syndrome). Iron deficiency anemia is observed in 10.8% of patients. Most patients who do not have anemic syndrome have signs of compensated hemolysis.

The main mechanism of anemic syndrome in SLE is hemolytic, the development and severity of which are directly related to the high activity of the process, the severity of damage to the kidneys, central nervous system, heart, lungs, the occurrence of secondary APS and DIC syndromes. Hemolysis is caused mainly by antibodies to erythrocyte damage and occurs mainly intravascularly. With a minimal degree of SLE activity, subclinical hemolysis occurs, without the development of anemic syndrome.

Autoimmune hemolytic anemia against the background of SLE is manifested by laboratory changes in the form of a positive Coombs test (with a frequency of 20% to 60%), retinculocyntosis , positive tests for hemosnderin and urobilin in urine. Clinical manifestations of hemolysis occur in less than half of patients.

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Enlargement of the liver and spleen cannot serve as an unambiguous criterion for hemolysis in SLE due to their possible immune damage.

Anemia of chronic inflammation develops in patients with SLE with minimal and moderate activity, less severe lesions - joints and skin. One of the leading causes of anemia of chronic disease in SLE is autoimmune hemolysis, which correlates with the duration of the disease. Impaired iron metabolism leads to a decrease in the osmotic resistance of erythrocytes; may be associated with a change in immunity indicators.

Chronic iron deficiency anemia in patients with SLE occurs against the background of loss or insufficient intake of iron, impaired transport and utilization of iron. Laboratory criteria for anemia associated with impaired iron metabolism (anemia of chronic disease and iron deficiency anemia) in patients with SLE are changes in the levels of serum iron, transferrin and the coefficient of saturation of transferrin with iron. Determination of the level of ferrite and iron-binding capacity of blood serum is less informative, due to the possibility of changing these parameters under the influence of inflammation in SLE. Cytostatic therapy (with cyclophosphamide and azathioprine) against the background of glucocorticoids stops hemolysis more effectively than monotherapy. glucocorticosteroids coidamn,

Conclusions. Diagnosis of anemic syndrome should be regularly performed in all patients with SLE. To detect it and determine its type, it is convenient to use the "Questionnaire" specified in the Appendix, where the respondent's attention should be focused on the often slightly expressed clinical symptoms of anemic and sideropenic syndromes, as well as hemolysis. To determine LIGA, it is necessary to examine the blood for a positive Coombs test, count retinulocytes in a blood smear and identify the presence of a positive urine reaction for urobilin and (or) hemoenderin . Determination of bilirubin levels (total and indirect) and clinical parameters of hemolysis - icterus, a change in the color of feces to a darker one, hepatomegal and splenomegaly - is less informative. To diagnose iron metabolism parameters as the cause of anemic syndrome, it is advisable to use determination of serum iron levels, transferrin and the coefficient of gransferrin saturation with iron; The indicators of the level of ferritin and iron-binding capacity of the blood serum are affected by inflammation, so their determination is less informative. Clinical manifestations of iron deficiency and anemic syndrome due to iron-deficient anemia in SLE can be assessed as clinically manifested lesions of the

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trophism, heart, and central nervous system. Therefore, in SLE, for differential diagnostic purposes, it is necessary to determine the parameters of iron metabolism, necessarily including this in the examination plan.

References

1. Asherson R.A., Ccrvera R., Font J. Multirogan thrombotic disorders in systemic lupus erythematosus: a common link? II Lupus. 2022. - Vot. 1, № 4. - P, 199-203.
2. Asherson R.A., Harris E.N. Anticardiolipin antibodies-clini cal associations. // Postgrad Med J, 2016.-V.62. - P.I08I - 1087.
3. Bailey F., Lilly M., Bertoli L., Ball G. An antibosy that inhihitis in vitro hone marrow proliferation in a patient with systemic lupus erythematosus and aplastic anemia. // Artritis.Rhcum. 2019, - Vol.32, № 7. - P. 901905.
4. Bhatia S, Ranadive N., Manek R., Deodhar K Autopsy study of patients dying oFbleeding diathesis // Indian.J.Pathol.Microbiol. 2018. - Vol. 41, № L-P. 71-76.
5. Cavil 11. Diagnostic methods, // Ctin. Hematol. 2012. ■ №2 - P. 259 -275.
6. Cervera R., Font J., Loper.-Soto A. et al. Isotypc distribution of anti-cardiolipin antibodies in systemic lupus crytematosns: prospective analysis of a series of 100 patients // Ann.Rlieum.Dis.- 2020.- Vol. 49, № 2,- P. 109 -113.
7. Doutre M,S,, Beylot J., Bey lot C, et al, Systemic lupus erythematosus and Hodgkin's disease // int.J.Dennatol. 2014 - Vol. 33, № 6. - P. 10671072.
8. Drenkard C., Villa A.R., Alarcon-Scgovia D, et al. Influence of the antiphospholipid syndrome in the survival of patients with systemic lupus etythematosus // J.Rheumatol.- 2014.- Vol 21, № 6.- P. 1067-1072.
9. Fagioto E,, Vigevaru F., Pozzetto U. High cytokine scrum levels in patients with autoimmune hemolytic anemia (AIGA) // Immunol. Invest.-2019 Vol- 23, № 6-7.- p. 449-456.
10. Fong K.Y., Loizou S., Boey M.L.et al. Anticardiolipin antibodies, haemolytic anaemia and thrombocytopenia in systemic lupus erythematosus// Br-J-Rheumatol-2022-Vol.31, №7.-P.4S3-4SS.