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FEATURES OF SYSTEMIC LUPUS ERYTHEMATOSUS IN CHILDREN

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Hematological parameters are of great importance in the diagnosis of SLE, especially in determining its activity and the risk of progression. The hematological manifestations observed in SLE can be represented by both true autoimmune phenomena of autoimmune hemolytic anemia (AHA), leukopenia (LP), and thrombocytopenia (TP), as well as cytopenic syndromes associated with the use of immunosuppressive drugs. Currently, it is known that hematological manifestations vary significantly in severity and often do not require specific treatment, with the exception of severe cytopenia, refractory to glucocorticoids (GC) [3]; at the same time, their significance as possible predictors of the further course of SLE has not been sufficiently studied.

One of the causes of anemia in SLE is anemia of chronic diseases (ACD), the cause of which is considered to be a disorders of iron metabolism in the macrophage system under the influence of inflammatory cytokines [2,4,5]. This type of anemia in SLE is much less studied, although the frequency of ACD in SLE varies from 11.9% to 37.1%. [1,3,5]. There is practically no information about the existence of iron deficiency anemia (IDA) in SLE, although this type of anemic syndrome can occur in these patients as well as in the general population. In single studies, it is indicated that changes in the level of serum ferritin and the content of bone marrow sideroblasts are practically not informative in the diagnosis of this type of anemia (Lila A.M., 2017).

Thus, the pathogenetic mechanisms of anemic syndrome in SLE are complex, while there are no clear clinical and laboratory criteria for it, and there are practically no data on the relationship between anemic syndrome and clinical manifestations of SLE [4,5].

Aim of study - to characterize the clinical and hematological signs of SLE in patients of different age groups.

Materials and methods. We examined 36 patients (30 girls and 6 boys) with SLE aged 6 to 18 years who were treated at the Tashkent State Medical University multidisciplinary clinic in 2020-2025. The diagnosis was established in accordance with the 2012 SLICC (Systemic Lupus International Collaborating Clinics) criteria [1]. A mandatory condition for the inclusion of patients in the study was their signed informed consent. The average age of the examined patients was 11,2±3,6 years.

The average time from the onset of the disease to the establishment of a diagnosis and the start of treatment was 6,3±0,7 months. At the time of the examination, the average duration of SLE in years was 8.7±2.8 (27,1±5,3 in months). All patients underwent a standard examination, which included an assessment of the activity of the disease according to the SLEDAI 2K index (Systemic Lupus Erythematosus Disease Activity Index-2K), immunological parameters: antinuclear factor, rheumatoid factor (RF), clinical and biochemical analyzes of blood, urine according to unified methods.

Results and discussion. Among the examined patients, there were various systemic signs of the disease. All patients had damage to the skin, joints, kidneys, heart, lungs. The frequency of joint damage in the examined patients was 82,0% (n=31). The most common were polyarthritis 60%

(n=28), skin lesions in the form of facial erythema 66% (n=25) (Fig. 1.), discoid lupus 50% (n=18) and photosensitivity 60% (n=21). Lung involvement was determined in 20 (58%) patients with SLE. Most often, X-ray or computed tomography of the lungs revealed infiltrates 25% (n=9), less often cavities in the lungs 10% (n=3). Respiratory system involvement included involvement of various lung regions: airways, parenchyma, vessels, pleura, and diaphragm.

Kidney damage was detected in 70% of SLE patients (n=31).

Heart damage was observed in 18 (50%) patients with SLE. In 10 (32%) patients, according to ECHO-CG, atherosclerotic changes in the aortic valve were determined.

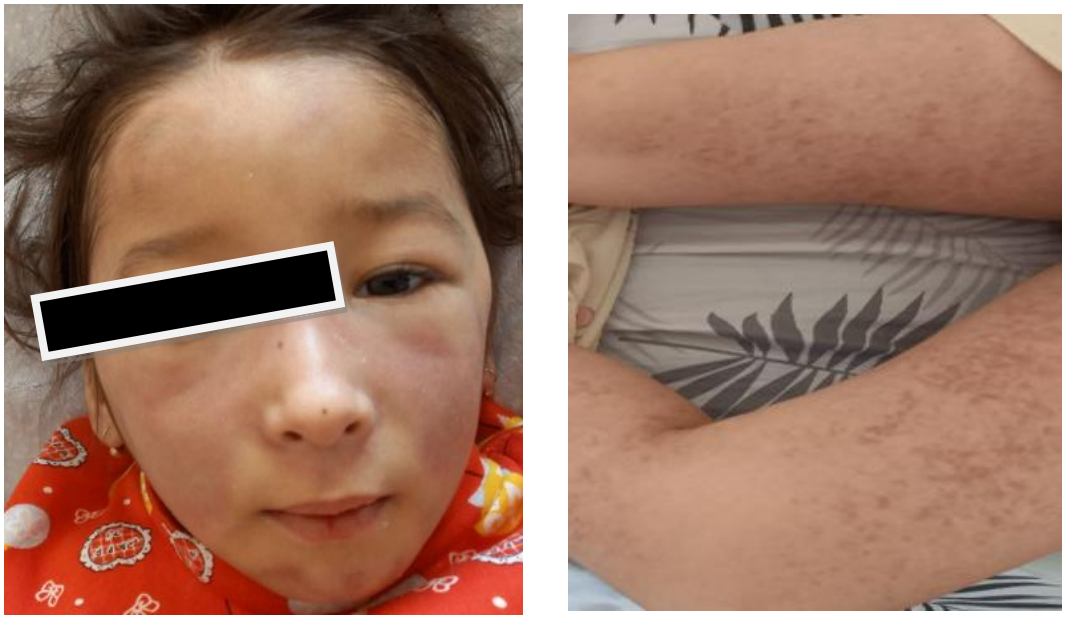

Fig. 1. Patient O. Skin manifestations of SLE. (Case report No. 319/56).

To compare ACD and IDA, patients were divided into 2 groups. Group 1 consisted of patients with ACD, group 2 patients with IDA. Of these, mild anemia was in 82% of patients, moderate - in 12%, severe - in 6%.

There were no signs of overt or covert bleeding. The groups were matched for sex, age, and disease activity. In group 1 with ACD, a significant decrease in reticulocytes was observed (possibly associated with hemolysis), high or normal ferritin levels, and anemia was hypochromic in nature with a normal level of serum iron. These data correlate with the data of other authors [3,5].

In the group with iron deficiency anemia (Group 2), there were mainly signs of a decrease in iron stores, a low level of ferritin, an increase in the total iron-binding capacity of serum with a low level of serum iron. Among patients of group 1, thrombocytopenia was significantly higher. Leukopenia occurred in both groups, but especially low rates were observed in patients of group 1. Some researchers regard leukopenia as a manifestation of an autoimmune process as a result of the production of antibodies to peripheral blood leukocytes, which correlates with the degree of SLE activity [4,5].

Other authors associate leukopenia with increased apoptosis of peripheral blood lymphocytes, morphological and genetic changes in neutrophils [2]. It is noteworthy that patients with severe leukopenia had a statistically significant prevalence of chronic SLE. A decrease in hematocrit was typical for patients in both groups, but significantly reduced in patients of group 2. This was possibly accompanied by hypoxia of various organs, since it is the red blood cells that normally carry oxygen

throughout the body, which worsened the condition of the patients. Polyarthritis and nephritis occurred in both groups, but significantly more often in patients with ACD in the form of active nephritis [1,3].

The examined groups were dominated by patients with chronic SLE (75.4% and 67.9%, respectively). Among the manifestations of SLE, lesions of the lungs (52.3%), kidneys (50.8%) and the nervous system (52.3%), occupied the central place in frequency, while lesions of the gastrointestinal tract (7%) were much less common. Kidney damage developed early (after 2.5 ± 2.3 months from the onset of general and/or local symptoms) and mainly in patients with ACD. Arthritis had a mono-oligoarticular character with a predominant lesion of the hand, large joints, sometimes it resembled the debut of RA. A decrease in hemoglobin level (110 g/l ≤ Hb < 120 g/l) was determined in 57% of patients, and a decrease in serum iron levels with normal hemoglobin levels in 18%.

Thus, a total iron deficiency state, including developed IDA, was observed in 93% of SLE patients. Among the clinical manifestations of SLE, polyarthritis, facial erythema, nephritis, alopecia, myocarditis, Raynaud's syndrome were more common in patients with ACD. ACD was characterized by a moderate decrease in hemoglobin (95±5.1 g/l versus 85±2.6 g/l), normocytosis, normo- or moderate hypochromia of erythrocytes, and a moderate increase in the number of reticulocytes.

Conclusion. Reduced serum iron levels and transferrin saturation are noted in both anemias, but the cause is different. If in IDA these changes are associated with absolute iron deficiency, then in ACD iron reserves are sufficient, but they cannot be utilized from the reticuloendothelial system.

An indicator of the state of the iron depot is the level of serum ferritin, which had normal or elevated values in ACD, while it was reduced in IDA. The severity of anemia correlated to some extent with the severity of the disease. Thus, IDA occurred in all children with stage III SLE activity. At the same time, it was of a severe and moderate nature, while with a lesser severity of the disease, mild anemia was more often observed.

Literature

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