IMMUNE STATUS IN HEMOGLOBINOPATHY

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Today, hemoglobinopathies are not limited to any particular region, they are widespread diseases around the world and represent a global public health problem. Hemoglobinopathies spread due to the migration of the population from endemic areas to countries where they were extremely rare among indigenous people.

Thalassemia is a disorder of the synthesis of one or more globin chains, a heterogeneous group of diseases in which the production of normal hemoglobin in the body is partially or completely inhibited and severe anemia develops [1-3]. Clinically important and most common types are a-thalassemia (D56) and β-thalassemia (D56.1). Globin β thalassemia with impaired synthesis of -chain β-called thalassemia. This type of thalassemia is more common than others. With a-thalassemia, the synthesis of a-chain is disturbed. Also described are cases of γ- and δ-thalassemia, in which the synthesis of globin chains of the same name is disturbed.

Thalassemia occurs in the Mediterranean basin, the Middle East, South and East Asia, the South Pacific and South China, with a carrier rate of 2 to 25%. More than 70% of newborns with sickle cell disease are born each year in sub-Saharan Africa [1,6].

Despite the lack of reliable information about the situation in many regions of the world, according to the latest information in the world, about 7% of the population is a carrier of hemoglobin disorder genes.

Today, hemoglobinopathies are not limited to a specific region, they are widespread throughout the world and therefore a global health problem. Hemoglobinopathies spread in connection with the migration of people from endemic areas to countries where it is very rare among the local population.

The purpose of the study

β-determining the ferritin level and cytokine status indicators of patients with thalassemia and analyzing the effectiveness of chelation therapy (XT).

Scientific news

For the first time, the relationship between cytokine status and ferritin levels and overall morbidity was investigated.

Materials and methods

In 200 patients with thalassemia registered at the Republican Specialized Hematology Scientific and Practical Medical Center, a study of the level of cytokines was conducted on the basis of RIGIATM together with the Institute of Immunology and Human Genomics. 60 of them are women and 140 are men.

Results and discussion

Since thalassemia is a hematological disorder, specialized care is provided in hematology facilities. It is known that there are 14 regions in Uzbekistan, in which 10-15 new cases of thalassemia are recorded every year. Lack of chelator therapy leads to hemosiderosis of internal organs (accumulation of excess iron) and death [5,7]. About 10-12 patients die each year before reaching adolescence. In recent years, the death rate has decreased due to chelator therapy. Thalassemia is most common in Surkhandarya, Kashkadarya, Samarkand and Bukhara regions.

Consanguineous marriages are known to increase the risk of developing thalassemia, and according to our data, consanguineous marriages are high in these areas.

Thalassemia is treated with chelation therapy, the main goal of which is to keep the body's iron concentration safe. Since 2015, the drug "Eksijad", which patients with thalassemia receive at the expense of state funds, has been supplied to our republic. At the place of residence or at RIGIATM, all patients are provided with the following medical care at the expense of the budget (i.e. free of charge): examination, basic therapy - transfusion of washed/thawed erythrocyte mass, additional therapy - vitamins, hormones, hepatoprotectors, symptomatic therapy and others), free hospitalization [4].

We analyzed the effectiveness of chelation therapy in patients with thalassemia in 2015-2020. A retrospective and report study and a comparison of the age table of patients with thalassemia before and after the introduction of chemotherapy showed that in the period after its introduction in the republic, the number of adolescents and adults who did not live to this age before was almost 2 doubled. The number of young children decreased by 3 times, the number of children of preschool and school age increased significantly, which also indicates a decrease in the death rate at this age.

Today, 36 patients over 17 years old are registered. Thus, the introduction of chelator therapy made it possible to increase the life expectancy of patients and reduce mortality. Before chelation therapy,

about 10-15 children with thalassemia died every year in the republic, in the years when chelation therapy was introduced, only 9 children died, and in 2021, 3 children died. While various infections were the cause of death, the total number of patients with thalassemia increased by 44 % in the years (2015-2022) when chemotherapy was introduced, indicating a decrease in mortality among them.

At the initial stage of diagnosis of thalassemia, continuous chelation therapy allows maintaining the level of iron in the body at a safe level, thereby preventing iron poisoning, preventing the development of hemosiderosis and, accordingly, disrupting the functions of internal organs. Without chelation therapy, almost all thalassemia patients develop severe hemosiderosis by the 5th to 6th year of life, with an average life expectancy of 6 to 8 years.

The main indicator of the development of hemosiderosis is the level of ferritin in the blood [8,9].

As our results showed before chemotherapy, serum ferritin level was 32870 ng/ml, i.e. more than hundreds of times. After 4 months of chelator therapy, this indicator decreased by 1.5 times and was 21708.1 ng/ml. At 8 months and 12 months after chemotherapy, ferritin levels decreased 2.5- and 4-fold, respectively, to 12905.9 and 7626.8 ng/ml ($p<0.05$).

Thus, chelator therapy showed its high efficiency, as well as the possibility of predicting the dynamics of ferritin levels in patients with thalassemia.

The increased susceptibility of patients with thalassemia to infections makes it necessary to study various aspects of the immune status of patients.

Thus, interleukin-2 (IL-2) indicators were on average 123.6 ± 114.1 IU / ml, and interleukin 6 (IL6) indicators were 241.4 ± 43.2 pg / ml

Normal indicators of IL-2 are 158-623 IU/ml, IL-6 - 3.40-5.90 pg/ml. Thus, the level of IL-2 increased 2-3 times compared to the norm, and IL-6 - hundreds of times. Perhaps this is due to hemosiderosis, which is confirmed by a significant increase in the level of ferritin. The obtained results indicate the need to study the relationship between hemosiderosis and IL-2 and IL-6.

In the study of interleukins in patients with thalassemia, an increase in the concentration of IL-2 and IL-6 in the blood serum was found, which indicates a shift in the production of interleukins to those that are anti-inflammatory.

Summary

The correlation between the level of ferritin and general morbidity was determined, which allows predicting the change of interleukin parameters

during chelation therapy. A significant increase in the amount of IL-2 and IL-6 in the blood serum of patients was found, which indicates a state of immunodeficiency.

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