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STATE OF THE IMMUNE SYSTEM AND THE DEVELOPMENT OF POST-TRANSFUSION COMPLICATIONS IN PATIENTS WITH HEMOPHILIA

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Abstract

The purpose of the study is the study the immune status of patients with hemophilia. A complex of immunological studies aimed at determining individual populations and immunoregulatory subpopulations of lymphocytes was carried out in 45 patients with hemophilia, of which 25 suffered from a severe form and 20 were diagnosed with an average form of hemophilia. 16 practically healthy men were examined as a control group. The research results show that hemophilia is accompanied by a violation of the immune status in patients, which manifests itself in changes in the cellular link (a sharp decrease in the total pool of T-lymphocytes and its subpopulation - TGF (theophylline-resistant) cells, with significantly high values of THF (theophylline-sensitive) cells and humoral immunity (increase in the content of B-lymphocytes, severe dysimmunoglobulinemia with a decrease in the content of immunoglobulin A and an increase in the level of immunoglobulins M and G). Data from studies of FAN (phagocytic activity of neutrophils) and CEC (circulating immune complexes) also confirm the fact of impaired immunoreactivity in patients with hemophilia: FAN values are reduced, increasing CEC.

Keywords: hemophilia, cellular, humoral immunity, immunoreactivity.

Introduction

Hemophilia is the most common form of hereditary coagulopathy - manifests itself in early childhood and is characterized by hemorrhages in the joints, and muscles and life-threatening bleeding into internal organs, which leads to a decrease in working capacity and early disability of patients [1,2,5,6]. It accounts for 68-78% of all cases of hereditary blood coagulation disorders. The birth rate of patients with hemophilia in each country is constant and according to WHO, the prevalence of this pathology in the world is 6-18 patients per 100,000 population. Multiple blood transfusions, inevitable in the treatment of hemophilia, lead to the breakdown of immunoregulatory mechanisms, and although, according to the literature, this aspect has several developments, the clinical significance of immunodeficiency in this category of patients remains unestablished [3,4,7,8,9]. Clarification of this issue is important for choosing the most rational treatment regimens for complications of hemophilia. In this regard, the study aims to study the immune status of patients with hemophilia.

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Materials and research methods

Under our supervision and treatment were 51 patients with hemophilia, of which hemophilia A was diagnosed in 37 and hemophilia B in 14 patients, which respectively amounted to 72.5% and 27.5%. All patients were registered at the NIIG and PC of the Ministry of Health of the Republic of Uzbekistan. Among the surveyed - all males aged 10 to 43 years. All subjects underwent immunological studies of cellular and humoral immunity. The data obtained as a result of the study were subjected to statistical processing using Student's criterion.

Research results

A complex of immunological studies aimed at determining individual populations and immunoregulatory subpopulations of lymphocytes was carried out in 45 patients with hemophilia, of which 25 suffered from a severe form and 20 were diagnosed with an average form of hemophilia. 16 practically healthy men were examined as a control group. As a result of the study of the content of the total pool of T-lymphocytes in the peripheral blood of patients with hemophilia during the period of recurrence of the disease, a decrease in both their relative and absolute numbers was revealed (52.8 ± 1.1% and 841 .6±36.2 in 1 µl) compared with the healthy group (respectively, 61.1±1.2% and 993.7±37.5 in 1 µl, $p<0.001$, in both cases). At the same time, in terms of the average value of T-lymphocytes in the blood of patients with hemophilia, a certain dependence of the level of cellular immunity on the form of the severity of the disease and, accordingly, on the volume and duration of transfusion -corrective therapy was revealed. Thus, in patients with moderate to severe form with relapses of the disease, a decrease in the content of T-cells (54.2±1.2%) was observed, in comparison with the control group (61.1±1.2% $p<0.001$) and more pronounced - in severe form (48.6±2.0%, $p<0.001$ compared with control. Intergroup differences were also revealed, indicating lower relative content of T-lymphocytes in the group of patients with severe hemophilia ($p<0.05$) compared with moderate hemophilia. subpopulations of T- lymphocytes revealed a significant increase in the number of theophylline-sensitive cells (TPCh) in the patients studied by us, amounting to 298.0 ± 25.2 per µl in absolute values and 18.7 ± 1.2% in relative values, compared with the group of healthy -227.7±24.1 in µl and 14.0±1.2%, respectively ($p<0.05$ for the first case and $p<0.01$ for the second). A more pronounced increase in THF was found in patients with severe hemophilia in comparison with the moderate form in relative terms ($p<0.05$). An increase in the level of THF, which has a suppressive effect, was accompanied by a decrease in the number of theophylline-resistant cells (TFR) that perform a helper function, both in absolute (604.1 ± 37.5 in µl) and in relative terms (37.9 ± 1.2%), in comparison with healthy people (725.4±35.9 per µl and 44.6±1.4%, respectively, $p<0.05$ and $p<0.01$ for each case). to the level of the healthy group, but with significance ($P<0.05$) differ in relative terms, amounting to 40.5±1.6%, against 44.6±1.4% in the control. Comparison of TGF values in severe hemophilia and a healthy group revealed a difference both in relative values (36.4±1.2% versus 44.6±1.4% in the healthy group, $p<0.001$) and absolute values (557 .7±33.7 per µl versus 725.4±35.9 per µl in healthy people, $p<0.001$). In addition, a significant difference was found between the two forms of hemophilia in terms of the relative TFR of 936.4±1.2% compared with 40.5±1.6%, $p<0.05$. Analysis of the parameters of humoral immunity in patients also indicates changes in immunoreactivity in the studied pathology. Thus, the content of the number of B-lymphocytes in the peripheral blood of patients was increased: in the general group of patients, it was 17.9 ± 1.4% in relative and 285.3 ± 28.5 per µl in absolute values (in healthy -

12.9 ±1.22 and 209.8±23.1 in µl, respectively, p<0.01 and p<0.05). In patients with severe hemophilia, these figures were 18.8±1.4% and 298.4±26.5 per 1 µl compared with healthy people (p< 0.01 and p<0.05, respectively). In the moderate form of the disease, the relative and absolute values of B-lymphocytes were, respectively, equal to 16.8±1.2% and 270.2±23.7 per 1 µl, which is significantly higher when compared with the healthy group in relative values (p <0.05). The level of immunoglobulin A in the blood serum of the studied contingent decreases both in the general group of patients and in various forms of hemophilia. The content of immunoglobulin A in the general group of patients under consideration is 172.2±3.2 mg%, which is significantly lower than in healthy people (181.6±1.4 mg%, P<0.01). The decrease in the level of immunoglobulin a occurs mainly due to its significant decrease in patients with severe hemophilia (170.6±1.6 mg%). Differences are significantly significant both when comparing this group of patients with healthy (181.6±1.4 mg%, p<0.001), and with indicators in moderate hemophilia (178.3±2.0 mg%, p<0, 01). The content of immunoglobulin M in blood serum significantly increases in all three groups of patients under consideration - 139.4±3.4 mg% in severe form, 132.8±2.4 mg% in moderate form, and 136.0± 4.2 mg% in the general group of patients (in the group of healthy individuals 122.5±1.8 mg%, p<0.001 in the first case and p<0.01 in the other two, respectively). However, when comparing the two considered forms of hemophilia, the difference between them was statistically insignificant. The content of immunoglobulin G in the studied patients was determined at a higher level than in healthy ones. So, in the general group, this indicator was 1642.4±28.2 mg%, in the group with a severe form - 2090.5±33.4 mg%, and the moderate form 1501.7±16.6 mg% (in the group of healthy individuals -1104.0±18.1 mg%, p<0.001 in all cases). Differences in the level of immunoglobulin were significantly significant. G in the compared groups of patients with severe and moderate hemophilia (p<0.001). The phagocytic activity of neutrophils (FAN) is reduced in all three groups. Thus, the level of FAN in the moderate form was 52.4 ± 1.8%, in the severe form -47.3 ± 1.6%, and in the general group of patients, it was 50.6 ± 1.1% (in the healthy group -59, 4±1.4%, p<0.01 in the first case, and p<0.001 in the other two). The defectiveness of phagocytosis is an important condition for increasing the content of pathogenic immune complexes (CIC). This is also confirmed by our research. Thus, an increase in the content of the CEC in patients with hemophilia was established: in a severe form of the disease, it amounted to 0.071±0.011 arb. units with moderate -0.043±0.009 arb. units and in general per group - 0.065±0.002 arb. units compared with the group of healthy -0.01±0.002 arb. units (p<0.001 in all cases). There is a significant difference in CEC values between groups of patients with severe and moderate hemophilia (p<0.05).

Discussion

Thus, the results of studies show that hemophilia is accompanied by a violation of the immune status in patients, which manifests itself in changes in the cellular even (a sharp decrease in the total pool of T-lymphocytes and its subpopulation - TGF cells, with significant high values of THF cells) and humoral immunity (increased the content of B-lymphocytes, severe dysimmunoglobulinemia with a decrease in the content of immunoglobulin A and an increase in the level of immunoglobulins M and G). These studies of FAN and CEC also expose the fact of impaired immunoreactivity in patients with hemophilia: FAN values are reduced, increasing CEC. It should be especially noted that the most profound changes in all parts of the immune system with pronounced suppressor activity are observed in severe hemophilia.

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Conclusions

Identified secondary immunodeficiency the state of hemophilia, in turn, affects its clinical variability, creating conditions for the development of autoimmune processes, the manifestations of which are: post-transfusion complications, resistance to specific therapy, the development of secondary rheumatoid syndrome in patients with hemophilia, etc.

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