FREQUENCY OF OCCURRENCE OF THROMBOCYTOPENIC PURPURA AND METHODS OF THEIR TREATMENT

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Abstract. Clinical cases of treatment of thrombotic thrombocytopenic purpura using multiple exchange plasmapheresis are presented. The difficulties of rapid diagnosis and the importance of early initiation of pathogenetically determined therapy are described. The tactics of using extracorporeal hemocorrection in the treatment of this disease are discussed.

Keywords: Treatment, moschkowitz disease, method, thrombotic thrombocytopenic purpura, plasmapheresis.

INTRODUCTION

This disease was first described by E. Moschcowitz in 1924 [1] and subsequently named after him. Moschkowitz disease or thrombotic thrombocytopenic purpura (TTP) (microangiopathic hemolytic anemia, thrombotic microangiothrombosis) is one of the types of thrombotic microangiopathy, which is based on systemic damage to small vessels by antigen-antibody complexes, causing endothelial proliferation, fibrinoid necrosis, formation of fibrin and hyaline thrombi [2].

MATERIALS AND METHODS

TTP is a rare disease. M. Noris et al. [4] note an incidence rate of 2–4 cases per 1 million population per year, and predominantly women suffer (the ratio of incidence of women to men is 3:2–5:2). People aged 30–50 years are most often susceptible to the disease.

The Oklahoma State Registry (USA, 2013) contains data on 4.46 cases of idiopathic TTP per 1 million residents per year [4]. Over time, there is no trend towards an increase in incidence. According to studies conducted in the USA in 1966–1991. [8, 9], the incidence was 3.7 cases per year per 1 million inhabitants. All researchers agree that TTP develops as an acute disease with a poor prognosis. The aggressive nature of the course of TTP requires the initiation of pathogenetically based therapy already in the first hours of the disease; in the absence of treatment, mortality approaches 90% [3].

The basis for the treatment of acquired TTP is the treatment regimen developed by J. George, taking into account the recommendations of the British Committee for Standards in Haematology, British Society for Haematology [4]. According to this scheme, basic therapy for TTP should be started as soon as possible, mainly on the 1st day of the disease. Therapy involving plasma exchange in a volume of 40–60 ml/kg body weight per day is pathogenetically justified [2]. If it is impossible to immediately begin plasma exchanges, it is recommended to carry out infusions of significant doses of fresh frozen plasma – FFP (25–30 ml/kg body weight per day) [4]. The advantage of plasma exchange, including in terms of improving the prognosis and reducing mortality in this group of patients, has been shown in a number of randomized studies, and is explained by the fact that plasma exchange allows the removal of a metalloprotease inhibitor, and FFP infusion only introduces this enzyme into the body from the outside.

The purpose of the work is to use the example of two cases from practice to describe the features of the clinical picture, variants of the course, diagnosis and treatment of such a severe and rare disease as TTP.

RESULTS AND DISCUSSION

Clinical observation No. 1

Bol'ny G., 37 years old. From the anamnesis: the first signs of the disease appeared 2 weeks later. after an acute respiratory viral infection and included weakness, pale skin, changes in behavior (unmotivated aggressiveness, mental

disorders), dizziness, and fever. He sought medical help at his place of residence, and examination revealed severe anemia (hemoglobin 65–70 g/l), thrombocytopenia (12–16 × 109/l), moderate hyperbilirubinemia (total bilirubin 40 μmol/l, mainly due to - direct fraction). A day after hospitalization, progression of neurological symptoms was noted - depression of consciousness up to stupor, right-sided hemiparesis; The appearance and increase of respiratory failure, which required the start of artificial pulmonary ventilation (ALV), and severe bronchorrhea with the release of a large amount of serous-hemorrhagic sputum, were also noted.

Clinical observation No. 2

Sick M., 39 years old. From the anamnesis: he noticed the first signs of the disease 1.5 months before hospitalization (weakness, headache, fever up to 38 °C). He was treated independently, after a short (about 2 weeks) improvement he noted the appearance of yellowness of the skin, pain in the lumbar region, and transient swelling of the legs. He sought medical help at his place of residence and was hospitalized in the infectious diseases department. The examination excluded acute viral hepatitis, leptospirosis, and HIV infection. 2 weeks after hospitalization, a sharp deterioration in condition was noted - impaired consciousness (stupor), progressive anemia (decrease in hemoglobin concentration to 65 g/l, red blood cells to 1.79 × 1012/l), thrombocytopenia (up to 25 × 109/l) , a moderate increase in the concentration of total bilirubin (up to 50 μmol/l) mainly due to the indirect fraction, LDH.

The described clinical cases have both a number of common features and significant differences. In both situations, TTP developed in men, which, according to the literature [2], is not typical for this disease and, accordingly, was a factor complicating diagnosis. The provoking factor in both situations was probably ARVI, which corresponds to the data published in the literature [3]. The clinical picture was not quite specific at the onset of the disease in both patients, but gradually progressed over 1–1.5 months, which was reflected in the appearance of most of the known

classical symptoms of TTP (microangiopathic hemolytic anemia, severe thrombocytopenia, neurological disorders, hemorrhagic syndrome, fever).

CONCLUSION

Thus, despite the fact that TTP is a severe and, in the absence of specific treatment, a highly lethal disease, with correct diagnosis and timely initiation of therapy, it is quite possible to achieve not only a clinical cure, but also long-term remission.

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