

tered in a unit dose of prednisolone - 1 - 2 mg per 1 kg weight. In the absence of the effect of receiving glucocorticoids in a dose within 1 - 4 months' (45) made splenectomy. In 37 of their number after splenectomy complete remission of the disease was achieved and AITP not recurred more. With 8 of them after splenectomy had recurrent disease. Such patients we administered rituximab 4 - 6, infusion of 375 mg / m² once a week. After that, all was in remission. Within two years he performed with rituximab maintenance therapy - 375 mg / m² - 2 of the introduction of 3 - 6 months. After 2 years, all remained in remission and rituximab canceled.

Attempts to use rituximab before (instead of) splenectomy in any case, not result to achieve stable remission. That corresponds to the literature - rituximab is effective only after the removal of the spleen. [4]

Recombinant thrombopoietin not often used, due to their high cost. Revoleyd (eltrombopag) was ordered for three patients. Dosing regimen was administered individually based on the number of platelets, the initial dose - 50 mg 1 time per day. If after 2-3 weeks of initial therapy the platelet count remained below the level required from a clinical point of view (50,000 / ml), the dose was increased to the maximum - 75 mg 1 time per day. Standard dose correction downward or increase was 25 mg per day. At the level of platelet 200000-400000 / l reduced dose. Enpleyt (romiplostim) was administered to patients one to three times a week by subcutaneous injection. Romiplostim initial dose was 1 mg / kg body weight. Romiplostim weekly dose was increased in increments of 1 .mu.g / kg body weight as long as the number of platelets in the patient did not achieve more than 50 × 10⁹ / L. All patients treated with both forms of recombinant TPO in patients receiving drugs platelet count normalized. After its cancellation AITP recurred.

Fisher-Evans syndrome Classic (combination AITP gemolitiche and autoimmune anemia) was diagnosed in two patients. In both cases the remission was achieved after splenectomy.

Deaths from AITP only 5 patients (3%) was diagnosed in 10 years, there has been a brain hemorrhage in all cases.

Conclusion. Thus, there currently are available treatments for advanced high AITP. With timely diagnosis of the disease and the appointment of adequate therapy FDI in the majority of cases of the disease prognosis - favorable.

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RESPIRATORY AND NEURAL SYSTEM DAMAGE CAUSED BY SYSTEMIC LUPUS ERYTHEMATOSUS

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Abstract A brief review of the literature devoted to the disorder of the respiratory and neural system in systemic lupus erythematosus (SLE). Characteristics of the most common pathology - pleuritis are disorder of the diaphragm, upper respiratory tract, pulmonary vessels. Particular attention is given to lupus pneumonitis and pulmonary (alveolar) hemorrhage. As an example, here is the case of the personal experience of the authors. At the young age of the patient at the acute stage of SLE leading clinical manifestation was caused by lung disease, manifested by acute respiratory failure, hemoptysis, anemia, and X-ray picture of bilateral interstitial and alveolar lesions of the lung tissue. Based on the identification of laboratory signs of active SLE the lupus lung damage of hemorrhagic alveolitis type was diagnosed. After treatment with glucocorticoids (pulse therapy and long-term reception of high doses orally), pulse therapy of cyclophosphamide, cascade plasma filtration the regression of lung manifestations of SLE has been achieved.

Key words: systemic lupus erythematosus (SLE), respiratory and neural system.

Systemic diseases of connective tissue (SDCT) appear as a heterogeneous group of various diseases, which are accompanied by a variety of changes inherent in both cellular and humoral immune response, and are combined with autoimmune phenomena [1, 4]. In recent years, along with polysystemic lesions of SLE at the onset it is often to occur as the oligosyndromic or monosyndromic disease, mainly affecting the organs of various systems: heart, lungs, kidneys, nervous system and etc. Nervous system damage may take the form of isolated central nervous system angiitis, epileptic syndrome, stroke, migraine headaches and other. With all the variety of manifestations of SDCT, which is articulate, hematological, neurological, nephrological, dermal, etc. the changes in the respiratory system, especially those that affect the pulmonary interstitium, occur in more than half of patients in different embodiments [2, 3, 4]. The complications due to interstitial pulmonary disease often appear to be leading in determining the prognosis of patients. X-ray examination revealed strengthening in interstitial lung pattern and its bullous deformation, whereby appears the picture of alternation of sclerosis foci and emphysema, resembles a honeycomb or "opal glass" also known as "cellular lung" or "honeycomb lung". The most informative of noninvasive diagnostic methods for lung lesions in recent years has become a computed tomography (CT-scan), which allows to evaluate the activity of inflammation and fibrosis, which is important for determining the effectiveness of therapy. Acute specific lesion of the pulmonary parenchyma is quite rare disease and usually develops in the highly active systemic lupus erythematosus (SLE). Pleural and pulmonary changes may forego arthralgia and dermal lesion. The lung hemorrhage may be the first and only manifestation of SLE, may be combined with other organ damage, or even outpace other systemic manifestations of the disease. The risk of the lung hemorrhage is not dependent on the duration of the disease. Patients with pulmonary hemorrhages frequently occur fever, hemoptysis, rapidly decreasing hematocrit, and developing of clinical and radiological signs of acute lupus pneumonitis with massive infiltration. CT images is a lot like acute lupus pneumonitis, but the classic symptoms of vasculitis may be missing. Acute parenchymal lung damage in SLE can be represented by an acute lupus pneumonitis and hemorrhagic alveolitis. Lupus pneumonitis manifests itself with the development of dyspnea, cough, pleuritic pain and fever, and also with the presence of unilateral or bilateral pulmonary infiltrates which can be radiographically defined. In all cases, it's necessary to carefully exclude other causes of the symptoms, such as pneumonia, pulmonary embolism, heart failure, which often could complicate the course of SLE. Hemorrhagic alveolitis is less common manifestation of SLE (less than 2%) than lupus pneumonitis, but potentially fatal, with a mortality rate up to 50-90%, which develops, usually in the presence of extrapulmonary SLE lesions and is often associated with high activity of lupus nephritis [1, 3, 4].

Modern therapy with high doses of methylprednisolone started no later than 48-72 hours after the first acute episodes allows to successfully arrest the interstitial changes in the lungs. In this regard we want to represent what we believe to be an interesting clinical case of patient with SLE disease onset.

Patient B., woman, 28 years old, moved to the rheumatological department of ARCH from city hospital with complaints of shortness of breath at light load, a dry cough in the evening and at night, pain in the joints of the hands and ankles, feeling of palpitation at ease increasing while walking accompanied with the pressing pain in the heart, temperature rise up to 37.6°C in the evening, swelling in the legs, general weakness, loss of appetite, alopecia. From anamnesis it is known that in February 2015 the headache, weakness and fever up to 37.6°C has appeared. She was treated for upper respiratory tract infections (URTI) for 10 days with no improvement. During this period, there were swelling on the face, legs, joined by a sore throat; body temperature has increased up to 39°C. Three lytic mixtures was injected. In the middle of March, the cough has amplified, the feeling of palpitations appears, body temperature stays up to 37.6 degrees high, joined by pain in the joints of hands, knees, ankles. She was examined by a rheumatologist in a private clinic and from March 20, 2015 she received 10 mg of prednisolone. With treatment the swelling and the pain in the joints has reduced but fever, cough and shortness of breath remained. On March 30 the deterioration of health came in the form of intense shortness of breath, cough, blood in sputum and the pain in the heart. She was taken to the city hospital, wherein the laboratory and X-ray examination was performed and revealed severe anemia, treated with single blood transfusion and injection of potassium and magnesium products. For further examination and treatment, she was transferred to the rheumatological department of ARCH.

On admission patient had subfebrile temperature up to 37.2-37.4°C, pale skin and mucous membranes, swelling throughout the body and joints of the hand, hemorrhagic rash on shins, BP - 110 and 70 mm Hg, pulse - 100 beats/min, dry cough, shortness of breath at ease up to 20 breaths per minute, the weakening of the respiratory noise in the middle and lower lung areas, mostly the right side.

A complete blood count (CBC) shows: severe anemia (Hb - 52 g/l), erythrocyte sedimentation rate (Westergren ESR) accelerated up to 100 mm/hour, moderate hypoproteinemia 61 g/l, increasing level of urea up to 9.5 mmol/l. Urinalysis (UA) shows: relative density - 1020, 5 RBCs per high power field (HPF), protein - 0.3 g/l; microscopic examination shows: RBCs - 19733 in 1 ml, WBCs - 1,067 in 1 ml. Immunogram shows: IgA - 4,3 g/l (normal 0,9-5,0), IgM - 4,4 g/l (normal 0,7-3,7), IgG - 26 g/l (normal 9,0-20,0), circulating immune complexes within normal limits, complement C3 - 72 (normal 75-135), C4 - 12.5 (normal 9-36), antinuclear factor (ANF) detected, antinuclear antibodies detected, antibodies to dsDNA are found (six times above normal), antibodies to phospholipids detected (half times above normal), anti-neutrophil cytoplasmic antibodies (ANCA) are

found. Lupus anticoagulant is not detected. Microscopic examination of sputum shows: RBCs up to 50 per high power field (HPF). CT scans of the chest was performed which shows: subtotal reduction of pneumatization of both lungs due to pronounced and diffuse changes in the pulmonary parenchyma, the "opal glass" alike and moderately expressed reticular changes, the bronchopulmonary pattern is diffusely strengthened and deformed. According to fibrobronchoscopy the mucosa of the nasal passages moderately bleeds on contact. Aspirate with the presence of blood. Bloody secret comes from all of the small bronchi, and distal parts of the lungs. In the middle third of the trachea, in interchondral space on the outer wall of intermediate bronchi on the right and on the front wall of the left primary bronchus is found 2-3 petechial formation, from which the blood is released when straining. The se endoscopic findings indicate vasculitis. The puncture renal biopsy performed which shows membranous-proliferative glomerulonephritis. Echocardiogram shows no signs of pulmonary hypertension. Contractive ability of the left ventricle myocardium preserved. Pericardial exudate up to 50 ml. CT signs of pulmonary edema, bilateral hydrothorax. In the light of prevalence of pulmonary and minimal urinary syndromes differential diagnosis of SLE with ANCA-associated vasculitis was conducted. Given the presence of arthritis, alopecia, hemorrhagic alveolitis, membranous-proliferative glomerulonephritis, anemia was diagnosed SLE. Performed the pulse therapy with methylprednisolone 1000 mg three times, cyclophosphamide 1000 mg single dose, subsequent dose of prednisolone 60 mg per day, and two sessions held cascade plasma filtration, symptomatic therapy. During treatment the pulmonary hemoptysis, fever, respiratory failure, edema syndrome, were arrested. Positive dynamics in the form recovery pneumatization, resolution of interstitial changes in the lungs. Persist the phenomenon of hemorrhagic vasculitis, minimal urinary syndrome (microscopic hematuria, proteinuria minor), moderate anemia, subfebrile body temperature.

In the present case, on the background of high clinical and laboratory activity of SLE the patient developed acute lung damage, manifested by acute respiratory failure, hemoptysis, increasing anemia, and X-ray picture of bilateral interstitial and alveolar lesions of the pulmonary parenchyma. The presence of pneumonia, as well as other possible causes of pulmonary hemorrhage was not confirmed. The developing of the lupus lung lesions by type of hemorrhagic alveolitis was diagnosed.

Clinically hemorrhagic alveolitis flows as acute lupus pneumonitis, but differs in the rapid development of respiratory failure, accompanied by a decrease in hemoglobin and hematocrit due to pulmonary hemorrhage. Intensity of clinically overt hemorrhagic syndrome varies from profuse hemoptysis to pulmonary hemorrhage. However, the absence of hemoptysis does not exclude the presence of hemorrhagic syndrome in the lungs when the patient developing acute respiratory failure and radiographic infiltrates, accompanied by a decrease in hematocrit. The proposed mechanism of damage during this complication - necrotizing mikroangiit due to the deposition of immune complexes and the development of apoptosis involving the alveolar capillaries and venules. The presence of granular deposits of immune complexes and complement, identified by transbronchial biopsy, helps to distinguish a hemorrhagic alveolitis of SLE from other causes of pulmonary hemorrhagic syndrome (microscopic polyangiitis, Wegener's granulomatosis, Goodpasture's syndrome). Besides the already mentioned, one of the common causes of lung hemorrhage of SLE is the antiphospholipid syndrome, which was not confirmed in this case [2, 4]. Recurrence of hemorrhagic alveolitis after the injection of high doses of corticosteroids, despite repeated courses of pulse therapy with prednisolone, is quite typical for this kind of lung lesions of SLE.

With the development of hemorrhagic alveolitis in patients with SLE, given the unfavorable prognosis of this complication it is recommended to early beginning of the aggressive therapy with glucocorticoids. The efficacy of cyclophosphamide in the presence of a hemorrhagic alveolitis seems not to be apparent. It is believed that the plasmapheresis can also be useful in acute cases, particularly in the case of the resistance to the therapy described above, although the effect on the survival improving was not observed [2, 4].

The refore, the acute parenchymal lung disease with hemorrhagic syndrome (hemorrhagic alveolitis) is a rare but prognostically unfavorable complication of SLE. At suspicion on its development the differential diagnosis of pneumonia, aspiration, thromboembolic complications, which can often complicate the course of SLE, should be carried out. Recommended an early start of aggressive immunosuppressive therapy, primarily with glucocorticoids.

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