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Infectious complications in a patient with systemic scleroderma with polyorganic lesions: a case report

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The paper presents a clinical case of subacute course of systemic scleroderma with polyorganic lesions of lungs (pneumofibrosis, pulmonary hypertension), heart (restrictive cardiomyopathy, atrial flutter), kidneys (nephritis), conjoined with severe infectious complications (septicemia, phlegmon of the soft tissues of the left leg).
Key words: *systemic scleroderma, polyorganic lesions of lungs, heart, kidneys, phlegmon of the soft tissues, sepsis.*

Інфекційні ускладнення у хворого на системну склеродермію з поліорганними ураженнями: клінічний випадок

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У статті представлено клінічний випадок підгострого перебігу системної склеродермії з поліорганними ураженнями легень (пневмофіброз, легенева гіпертензія), серця (рестриктивна кардіоміопатія, тріпотіння передсердь), нирок (нефрит) з приєднанням тяжких інфекційних ускладнень (септицемія, флегмона м'яких тканин лівої ноги).
Ключові слова: *системна склеродермія, поліорганні ураження легень, серця, нирок, флегмона м'яких тканин, сепсис.*

Инфекционные осложнения у больного системной склеродермией с полиорганными поражениями: клинический случай

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В статье представлен клинический случай подострого течения системной склеродермии с полиорганными поражениями легких (пневмофиброз, легочная гипертензия), сердца (рестриктивная кардиомиопатия, трепетание предсердий), почек (нефрит) с присоединением тяжелых инфекционных осложнений (септицемия, флегмона мягких тканей левой нижней конечности).
Ключевые слова: *системная склеродермия, полиорганные поражения легких, сердца, почек, флегмона мягких тканей, сепсис.*

Systemic scleroderma (SSD) is an autoimmune connective tissue disease with characteristic lesions of the skin, blood vessels, musculoskeletal system, internal organs (kidneys, heart, lungs, digestive tract), based on impaired microcirculation, inflammation and generalized fibrosis. This disease is widespread worldwide. The average incidence is estimated to be 12–14 cases per 1 million population. Usually, the disease affects people of 30–40 years age group; it is by 4–8 times more prevalent in women than in men (7: 1 ratio) [3].

At the preliminary stage of diagnostics, the triad of the initial signs of the disease is especially important: Raynaud's syndrome, characteristic skin lesions, joint syndrome. Subsequently, multiple organs and tissues are involved in the process. Visceral lesions in SSD are quite diverse and determine the nature of the clinical progress of the disease and its prognosis. Cases are occasionally reported in the literature, describing unusual variants of the clinical progress of SSD and its complications [2, 7]. Cardiovascular lesions are one of the major visceral manifestations of SSD, with the involvement of the pericardium and vessels of all calibers; it is often a combined lesion.

Two types of lung lesions are characteristic:

– diffuse pneumosclerosis, mainly of basal parts, sometimes with cystic alteration, fibrotic alveolitis;

– isolated pulmonary hypertension or in combination with diffuse pneumosclerosis.

Lesions of all parts of the digestive tract are possible; esophageal lesions, intestinal lesions with impaired absorption and signs of obstruction, as well as ulcerative lesions are more often found. Renal lesions, diagnosed in 10-20% of patients, are one of the adverse factors affecting the survival of SSD patients.

Infectious complications are one of the major threats to patients with systemic connective tissue disease (SCTD), along with cardiovascular and oncological diseases [2,4]. Based on retrospective analysis, it has been established that the incidence of comorbid infections in the in-patients with rheumatic diseases was 9.7%; with predominant damage to respiratory organs (44%), urinary tract (29.2%), skin and soft tissues (18.9%). Comorbid infections were the most common in patients with systemic lupus erythematosus (SLE) (28.4%) and rheumatoid arthritis (13.2%). It has been established that about 50% patients with SLE suffer serious episodes of infection during their illness [1]. Infections account for 2-9% of all causes of mortality in patients with systemic scleroderma [8]. Cases of serious infections (pneumonia, sepsis, bacterial arthritis, skin and soft tissues lesions, etc.) have been recorded, including those with fatal outcomes.

Rheumatic diseases, including systemic connective tissue diseases, often result from secondary immunodeficiency. The use of immunosuppressive agents (glucocorticoids and cytostatics) in such patients, on the one hand, is necessary, since it is impossible to reduce the activity of the autoimmune process without them, and on the other side, strengthening of the existing immunodeficiency increases the possibility for infection development. One of the side effects of NSAIDs and cytostatics may be neutropenia, agranulocytosis, which is associated with a high risk of bacterial infection. Bacterial (staphylococcal, streptococcal, etc.) infections are predominant against the prevalent defects of humoral immunity (B-link). [1, 2, 9].

The urgency of this problem can be explained by the complexity of elucidating the primary cause of the disease, as the systemic response of the body can be caused by both infectious and non-infectious processes; complexity of interpretation of changes of physical and laboratory data (fever, tachycardia, tachypnea, abnormal leucoformula) and choice of tactics of management of patients with diffuse connective tissue diseases and sepsis. Presence of both similar symptoms associated with the active process in rheumatic diseases and immunosuppressive therapy, which is a leading risk factor for infection and concealment of its clinical symptoms, significantly complicate the diagnosis [1, 4, 10].

The problem of differential diagnosis of systemic infectious processes and rheumatic diseases occurring with high activity is extremely urgent. We report a case that we believe may be of particular clinical interest.

The 26-year-old male patient Z., presented with systemic scleroderma since 2016 when the signs of Raynaud's syndrome (pallor, cyanosis, hyperemia of the hands and feet) appeared, followed by thickened swelling of the skin, hyperpigmentation of the hands, face, telangiectasia on the face, pain and restriction of movement in the joints of the hands.

In the rheumatology unit after the examination the following diagnosis was made: SSD, diffuse form, subacute course, activity of type II, with skin lesions: induration, edema, hyperpigmentation; blood vessels: Raynaud's syndrome; lungs: basal pneumosclerosis;

digestive tract: reflux esophagitis. Treatment: 16 mg methylprednisolone, endoxane, vasoprostan, dipyridamole.

The patient recovered with improvement in health condition and lab tests after therapy. It was recommended to decline the dose of methylprednisolone and continue therapy with endoxane.

The patient's health condition worsened in 2019 after exposure to cold that was manifested by fatigue, shortness of breath, pain and swelling of the lower extremities, body temperature rised to 37.3 °C–38.5 °C. The patient was hospitalized in the rheumatology unit with subsequent transfer to surgical unit.

The condition of the patient at hospitalization was severe. The position of the body was forced because of pain in the lower extremities. The skin of the face, neck, trunk, limbs was thickened, hyperpigmented, dry, microstomia was apparent. Diffuse cyanosis, respiratory rate 22 breaths per minute. In the lungs, scattered dry wheezing associated with harsh breathing, crepitation on both sides at the inferior angle of the scapula. Heart rate 120 beats per minute, arrhythmic; blood pressure 80/40 mm Hg. Left border of the heart at the anterior axillary line, right border of the heart at the parasternal line. Heart activity was arrhythmic, tones were weakened, systolic murmur at the apex. The abdomen was soft, not tender. The liver protruded 1.5–2.0 cm below the costal arch. Swelling and hyperemia of the left shin and thigh; fluctuation in the middle third of the thigh.

On examination: CBC: RBC – $1{,}38\times10^{12}$/L, WBC – $22{,}3\times10^{9}$/L, haemoglobin – 27 g/L, platelet count – 5 22×10^{9}/L, ESR – 75 мм/год., stabs– 12%, lymphocytes – 4%.

Urinalysis: specific gravity – 1014, protein – 0.22 , WBC – 2–4 FOV, RBC – 1–2 FOV, daily urine – 350–400 ml.

Biochemical blood test showed total protein – 47.9 g/L, urea – 6.8 mmol/L, creatinine – 64 μmol/L, total bilirubin – 7.1 mmol/L, ALAT – 31 mmol/L, ASAT – 27 mmol/L, K – 3 mmol/l, Na – 130 mmol/L, total – 2.4 mmol/L, fibrinogen – 6.66 g/L, prothrombin index – 129%, INR – 0.77. Blood culture (2 times) revealed *Staphylococcus aureus.*

Chest and abdomen spiral computed tomography showed interstitial diffuse pneumosclerosis, fluid levels in the pleural sinuses, heart borders were widened; spleen: smooth contour, homogenous structure, enlarged in size measuring 130×97 mm. Conclusion: cardiomyopathy, bilateral exudative pleurisy, splenomegaly.

Echocardiography: dilatation of the heart chambers, low contractile capacity of the left ventricular (LV) myocardium, EF 31%. Mitral regurgitation of grade 2–3. Tricuspid valve regurgitation. Pulmonary hypertension, systolic pressure in the pulmonary artery 35 mm Hg.

ECG: atrial flutter 2:1; 3:1. Deviation of electrical axis of heart to the left. Frequent single and paired ventricular extrasystole. Block of the anterior left branch of the bundle of His. LV hypertrophy with the signs of overload and side wall ischemia.

Fibrogastroscopy (FGS) revealed erythematous gastropathy. Rectoromanoscopy showed ulcerative colitis.

Clinical diagnosis was made**:** systemic scleroderma, subacute course, activity III with skin lesions: induration, swelling, hyperpigmentation; blood vessels: Raynaud's syndrome, pulmonary hypertension I stage; cardiac lesions: restrictive cardiomyopathy, mitral regurgitation of grade 2-3, tricuspid valve regurgitation II; persistent atrial flutter, irregular (2:1; 3:1), tachystolic option, CHA2DS2VASc score1, HAS BLED score 1; ventricular extrasystole. CH IIA with reduced left ventricular ejection fraction (EF – 31%) FC III; digestive tract: reflux esophagitis, chronic gastroduodenitis at the stage of moderate exacerbation, ulcerative colitis; lungs: pneumofibrosis RD I, kidneys: sclerodermal nephropathy, CKD I.

Complications: staphylococcal sepsis, phlegmon of the soft tissues of the left lower extremity. Chronic posthemorrhagic severe anemia.

In the surgical unit, a wide dissection and drainage of the affected area was made. Antibacterial therapy (vancomycin, amikacin), intravenous Solu-Medrol, transfusion of fresh frozen plasma, erythrocyte mass, rheopolyglukin, pantoprazole, amiodarone was started. After therapy the patient recovered with positive dynamics, improvement in health condition and clinical and laboratory results. The patient was discharged in satisfactory condition with recommendation to continue antibiotic therapy with levofloxacin for up to one month.

The present clinical case clearly demonstrates a severe subacute course with polyorganic lesions of lungs, heart, kidneys, digestive tract. Immunodeficiency was accompanied by severe infectious complications, particularly, septicemia, soft tissue phlegmon, with complex pathogenesis that required prolonged intensive care.

CONCLUSIONS

1. The problem of differential diagnosis of systemic infectious processes and rheumatic diseases occurring with high activity is extremely urgent. The peculiarity of the clinical course of purulent-septic complications in patients with systemic connective tissue diseases is areactivity, obliteration of common manifestations of infection, the severe course, the unfavorable prognosis.

2. When supervising patients with rheumatic diseases, attention should be paid to the presence of risk factors in which the likelihood of the development of comorbid infections is increased, and it is important to identify early symptoms of infectious complication.

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