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CLINICAL FEATURES OF SJÖGREN'S SYNDROME ASSOCIATED WITH VASCULITIS

<https://doi.org/10.5281/zenodo.18368224>**Miraxmedova Khilola Tukhtasinovna****Mukhsimova Nilyufar Rakhimjonovna****Abduziyoyev Azimjon Dilshod ugli***mukhsimovanilufar@gmail.com**Tashkent state medical university*

Abstract: *This article presents the clinical course, diagnostic findings, and treatment process of a 29-year-old female patient with primary Sjögren's syndrome. The patient's complaints included dry mouth and eyes, multiple dental caries, joint pain, fatigue, purpuric skin rashes, and vaginal dryness — symptoms characteristic of this autoimmune disorder. Laboratory investigations revealed elevated inflammatory markers, positive rheumatoid factor, and the presence of hemorrhagic vasculitis. Due to insufficient response to standard therapy (prednisolone and hydroxychloroquine), the patient was initiated on biological treatment with Rituximab. This article analyzes the patient's clinical response to therapy, potential complications, and follow-up dynamics.*

Keywords. *Sjögren's syndrome; autoimmune disease; xerophthalmia; xerostomia; hemorrhagic vasculitis; biological therapy; Rituximab; immunological markers; clinical follow-up.*

ОСОБЕННОСТИ ТЕЧЕНИЯ СИНДРОМА ШЕГРЕНА В СОЧЕТАНИИ С ВАСКУЛИТОМ

Мирахмедова Хилола Тухтасиновна, Мухсимова Нилуфар Рахимжоновна,**Абдузиев Азимжон Дилшод угли***Ташкентский государственный медицинский университет*

Аннотация: *В данной статье представлено клиническое течение, диагностические показатели и процесс лечения у 29-летней пациентки с первичным синдромом Шегрена. Жалобы пациентки включали сухость полости рта и глаз, множественный кариес, боли в суставах, утомляемость, кожные высыпания в виде пурпуры, а также вагинальную сухость — симптомы, характерные для данного аутоиммунного заболевания. Лабораторные исследования выявили повышение маркеров воспаления, положительный ревматоидный фактор и наличие геморрагического васкулита. Из-за недостаточной эффективности стандартной терапии (преднизолон и гидроксихлорохин) пациентке была назначена биологическая терапия*

ритуксимабом. В статье проанализированы клинический ответ на лечение, возможные осложнения и динамические данные наблюдения.

Ключевые слова. Синдром Шегрена; аутоиммунное заболевание; ксерофтальмия; ксеростомия; геморрагический васкулит; биологическая терапия; ритуксимаб; иммунологические маркеры; клиническое наблюдение.

**SHEGREN SINDROMINING VASKULIT BILAN BIRGA KECHISHIDA DAVOLASH
TAMOYILLARI**

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Miraxmedova Khilola Tukhtasinovna

Mukhsimova Nilyufar Rakhimjonovna

Abduziyoyev Azimjon Dilshod ugli

Toshkent davlat tibbiyot universiteti

Annotatsiya: Ushbu maqolada 29 yoshli ayol bemorda kuzatilgan birlamchi Shegren sindromining klinik kechishi, diagnostik tahlil ko'rsatkichlari va davolanish jarayoni yoritilgan. Bemorning shikoyatlari og'iz va ko'z qurishi, tish kariyeslari, bo'g'im og'riqlari, charchoq, terida purpura ko'rinishidagi toshmalar va vaginal qurishlik kabi kasallikka xos simptomlardan iborat bo'ldi. Laborator tekshiruvlarda yallig'lanish markerlarining oshishi, RF musbatligi hamda gemorragik vaskulit mavjudligi aniqlangan. Standart davo (prednizolon va gidroksikloroxin) yetarli samara bermaganligi sababli bemorga Rituximab bilan biologik terapiya qo'llandi. Maqolada ushbu davo bosqichiga bemorning klinik javobi, asoratlari va kuzatuv dinamikasi tahlil qilingan.

Kalit so'zlar. Shegren sindromi; autoimmun kasallik; kseroftalmiya; kserostomiya; gemorragik vaskulit; biologik terapiya; Rituksimab; immunologik markerlar; klinik kuzatuv.

Sjögren's disease (SjD), previously known as Sjögren's syndrome, is a chronic autoimmune disease that primarily affects the body's exocrine glands, especially the lacrimal glands and salivary glands. The most common symptoms are dry mouth and dry eyes; however, the disease often also seriously affects other organs such as the lungs, kidneys, and the nervous system.

Global Prevalence

Worldwide, the prevalence of Sjögren's syndrome varies significantly across different sources. According to research findings:

The global prevalence of the disease is generally estimated to range from 0.1% to 1.0%.

Some epidemiological studies have reported figures ranging from 0.1% to 4.8%.

According to a 2025 meta-analysis, the global prevalence of Sjögren's syndrome was estimated to be approximately 13%.

These differences are explained by variations in diagnostic criteria and the variability of clinical manifestations.

Why Is the Disease Relevant?

Sjögren's syndrome is considered a relevant disease due to several clinical and social reasons:

1. Autoimmune nature: it can affect multiple organs beyond the glands.
2. Chronic course: a long-lasting inflammatory process that persists for years.
3. Delayed diagnosis: often identified late due to nonspecific symptoms.
4. Impact on quality of life: pain, fatigue, sleep disturbances, and reduced social activity.
5. Extraglandular involvement: vasculitis, kidney involvement, pulmonary pathology, and nervous system damage.

Factors Leading to Early Disability

Sjögren's syndrome contributes to the development of early disability due to the following factors:

- Ineffectively treated inflammatory process.
- Persistent joint pain and muscle weakness.
- Decreased quality of vision (due to dry eyes and keratoconjunctivitis).
- Chronic fatigue syndrome.
- Tooth loss and difficulty eating as a result of dry mouth.
- Vasculitis and involvement of internal organs.

Objective. Clinical observation of treatment outcomes in a patient with Sjögren's syndrome.

Patient: E.H.M

Age: 29 years

Sex: Female

Date of birth: 24.02.1996

Occupation: Housewife

Duration of disease: 5–6 years

Duration of complaints: Gradually worsening over the years

Complaints

The patient consulted a physician with the following symptoms that have persisted for 5–6 years: dry mouth (xerostomia) — difficulty swallowing dry food, frequent need to drink water, waking up at night due to dry mouth. Dry eyes (xerophthalmia) — burning sensation, blurred vision, thick mucoid discharge from the eyes. Dental problems — multiple caries, enamel erosion, loss of some teeth. Rashes on the legs — erythematous, reddened and purpuric spots on the skin, characteristic of autoimmune inflammation of the skin. Joint and muscle pain — especially in the knee area, with morning stiffness. General fatigue and weakness. Vaginal dryness — discomfort during sexual intercourse, dyspareunia.

Anamnesis vitae

The patient E.H.M., born in 1996, is a housewife. She married in 2015. Her reproductive history includes several unfavorable perinatal events. In 2018, her first pregnancy ended in a spontaneous abortion at 7–8 weeks. In 2020, a male child was born, but the child died at the age of 6 months (cause of death unknown).

In March 2025, the patient gave birth again. This newborn was diagnosed with congenital extrasystole. These facts indicate the presence of perinatal risks in the patient that may be associated with an autoimmune process, including the potential association of Sjögren's syndrome–specific autoantibodies (anti-Ro/SSA, anti-La/SSB) with pregnancy-related complications.

Physical examination findings

Skin: purpuric elements on the legs, consistent with hemorrhagic vasculitis.

Cardiac and pulmonary auscultation: no abnormalities detected (confirmed by echocardiography).

Parotid glands: no enlargement noted.

Body temperature: within normal limits.

Laboratory and instrumental data:

• General urinalysis (29.10.2025):

Volume – 70.0 ml*; Color – yellow; Clarity – clear; Specific gravity – 1010; Protein – 0.033 g/L; Squamous epithelium – 13–15; Transitional epithelium – 12–13; Renal epithelium – 10–11; Leukocytes – 13–14; Erythrocytes: altered – 0–1; unaltered – 0–1; Mucus – ++.

• Rheumatic tests (29.10.2025):

C-reactive protein – 25 mg/L [0–12]; Rheumatoid factor – 25 IU/ml (0–12); Antistreptolysin-O – 650 IU/ml (>400).

• Coagulogram (9 parameters) (29.10.2025):

Hematocrit – 38% [Women 38–42%; Men 49–58%]; Thrombin time (TT) – 23.2 s [up to 30 s]; Plasma heparin tolerance – 5–30 min [3–7]; Fibrinogen – 353 mg/dl [200–400 mg/dl]; Prothrombin time/index – 14.1 s / 78% [10–14 s; 75–140%]; INR (international normalized ratio) – 1.11 [0.8–1.25]; Activated partial thromboplastin time (aPTT) – 26.3 s [25–36 s]; Ethanol test – negative [negative]; Thrombotest – V [grade V].

• Biochemical blood analysis (29.10.2025):

Urea – 4.2 mmol/L [2.5–8.3]; Creatinine – 81.3 μ mol/L [Men 44–115; Women 44–97]; ALT – 24 U/L [<40]; AST – 13 U/L [<35].

• Complete blood count (extended) (29.10.2025):

Hemoglobin (HGB) – 100 g/L [Men: 130–160; Women: 120–140];

Erythrocytes (RBC) – 3.6×10^{12} /L [Men: 4.0–5.0; Women: 3.9–4.7];

Mean corpuscular volume (MCV) – 88 fL [80–100];

Platelets (PLT) – 218×10^9 /L [180–320];

Leukocytes (WBC) – 4.5×10^9 /L [4.0–9.0];

Neutrophils – 76%; Lymphocytes – 20%; Monocytes – 4%;

Erythrocyte sedimentation rate (ESR) – 20 mm/h [Men: 2–10; Women: 2–15].

- Liver and gallbladder ultrasound (24.10.2025):

Conclusion: Echo signs of changes in the liver parenchyma.

- Kidneys and adrenal glands ultrasound (24.10.2025):

Additional data: Bladder volume – 200 ml, wall thickness – 6.7 mm, intravesical fluid – turbid.

Conclusion: Echo signs of bilateral pyelonephritis, cystitis.

- Echocardiography (24.10.2025):

Conclusion: Left ventricular cavity not dilated: EDD – 4.0 cm, EDV – 74.0 ml, EF – 67.0%. Left atrium – 2.9 cm. Right heart chambers not dilated. Tricuspid valve – no structural changes. Mitral valve – no structural changes. Additional thickened chordae. Aortic diameter – 2.8 cm. Aortic valve – no structural changes. Right atrium diameter – 2.0 cm. Left ventricular walls normokinetic. IVS – 0.8 cm. Posterior wall of LV – 0.9 cm. LV myocardial mass – 103 g. Doppler EchoCG: no pathological flows. Pulmonary artery systolic pressure – 18 mmHg.

Conclusion: No congenital or acquired heart defects detected. Global contractility of the left ventricular myocardium is normal. No free fluid in the pericardial or pleural sinuses.

Primary health care results:

Diagnosis:

Primary diagnosis: Hemorrhagic vasculitis with skin involvement.

ICD-10: Allergic purpura D69.0

Recommendations:

Rituximab (Maball 500) 1000 mg on day 1, 1000 mg on day 14.

Prednisolone 90 mg → 60 mg → 30 mg with 0.9% sodium chloride 100 ml IV drip for 3 days; after completion, Prednisolone 5 mg, 1 tablet at 07:00 for 3 months orally (Hungary).

Plaquenil 200 mg, 1 tablet once daily at 19:00 after meals, long term.

Renalof, 1 capsule 3 times daily before meals for 15 days.

Devical, 1 tablet twice daily at 14:00 and 19:00 with meals for 2 months.

Vitamin D 5000 IU, 1 tablet once daily in the morning after meals for 2 months.

Reosorbilact 200 ml IV drip for 3 days; from day 4, Curantil 25 mg, 1 tablet 3 times daily after meals for 2 months.

Follow-up by a rheumatologist at the place of residence.

Results obtained after treatment

The patient's general condition was assessed as satisfactory after completion of the treatment course. During the first 3–4 days after the Rituximab injection, the patient experienced generalized body pain. These symptoms are explained by mild infusion-related reactions, and the patient was able to control the pain by taking Ibuprofen.

Against the background of treatment, the rashes and itching characteristic of hemorrhagic vasculitis on the legs completely regressed. This confirms the positive effect of biological therapy on the vascular inflammatory component. However, the main glandular manifestations of Sjögren's syndrome — dry mouth (xerostomia) and dry eyes (xerophthalmia) — did not decrease significantly. This condition may be associated with the chronic autoimmune nature of the disease and irreversible damage to the exocrine glands.

Since Sjögren's syndrome is a chronic disease, the patient should undergo preventive follow-up by a rheumatologist at least 1–2 times a year and repeat treatment courses as needed. This approach helps control disease activity, prevent new complications, and ensure early detection of glandular and extraglandular involvement.

Conclusion. Sjögren's syndrome is a long-lasting autoimmune disease, and its course in combination with vasculitis increases the risk of complications. Therefore, regular follow-up by a rheumatologist (1–2 times per year), laboratory monitoring, and repeated treatment courses when necessary are recommended. Early diagnosis, appropriately selected therapy, and continuous clinical observation play a crucial role in preventing severe complications of this disease.

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