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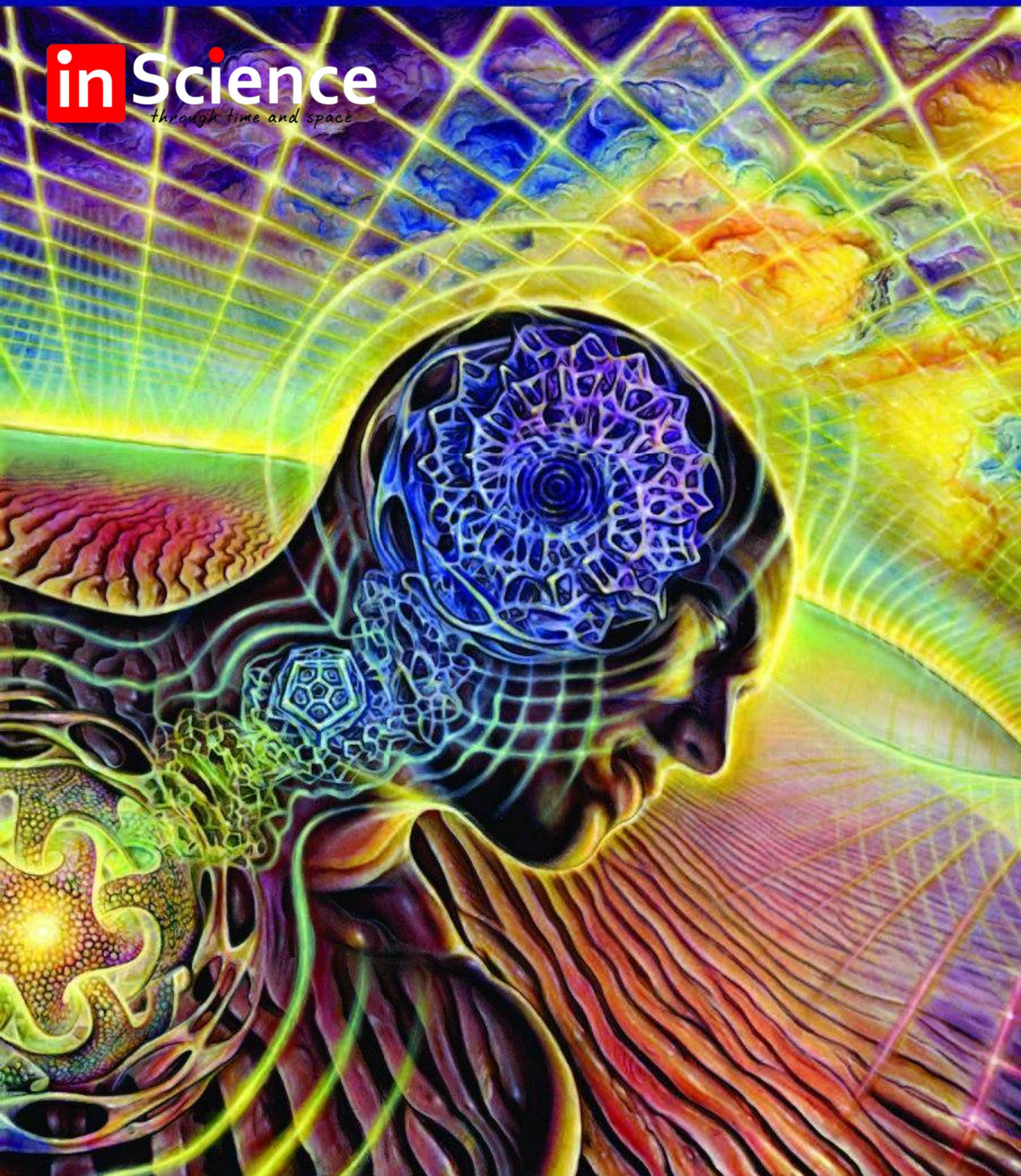

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ИННОВАЦИОННЫЙ ПОДХОД И ПЕРСПЕКТИВЫ СОВРЕМЕННОЙ СТОМАТОЛОГИИ И ЧЕЛЮСТНО-ЛИЦЕВОЙ ХИРУРГИИ

МАТЕРИАЛЫ
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ИННОВАЦИОННЫЙ ПОДХОД И ПЕРСПЕКТИВЫ СОВРЕМЕННОЙ СТОМАТОЛОГИИ И ЧЕЛЮСТНО-ЛИЦЕВОЙ ХИРУРГИИ
МАТЕРИАЛЫ МЕЖДУНАРОДНОЙ НАУЧНО-ПРАКТИЧЕСКОЙ КОНФЕРЕНЦИИ

Верстка: Хуршид Мирзахмедов

ХАРАКТЕРИСТИКА ПОРАЖЕНИЯ ЛОР-ОРГАНОВ ПРИ ГРАНУЛЕМАТОЗЕ ВЕГЕНЕРА

УЛУГБЕК САЙДАКРАМОВИЧ ХАСАНОВ,
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АННОТАЦИЯ

Болезнь Вегенера, некротизирующая гранулема верхних дыхательных путей с нефритом) впервые описана немецким патологоанатомом Ф. Вегенером в 1936, 1939 гг. [1-5]. Заболевание из группы системных васкулитов, характеризующееся гранулематозным поражением, прежде всего, верхних дыхательных путей, легких и почек. Постепенно заболевание переходит во вторую стадию, когда присоединяются признаки генерализации процесса: поражение внутренних органов (легких, печени, селезенки, почек), постоянная лихорадка, похудание. Третья стадия, терминальная, характеризуется признаками легочно-сердечной и почечной недостаточности. Помимо классической формы, гранулематоз Вегенера может протекать по «молниеносному» или «безголовому» типу. Диагноз ставится на основании типичной клинической картины. Из лабораторных показателей характерны высокая СОЭ (40-60 мм/ч), нормохромная анемия, у 50% больных выявляется ревматоидный фактор. Риноскопия важна. При гистологическом исследовании биоптата слизистой оболочки носа обычно выявляют васкулит мелких сосудов, но наиболее характерным диагностическим признаком являются гранулемы. Гранулема носа (лица) злокачественная (срединная, фатальная) характеризуется поражением носа, мягких тканей лица, полости рта, твердого неба с развитием обширных язвенно-некротических дефектов. Внутренние органы обычно не поражаются. За период с 2020 по 2021 г. мы наблюдали 3 случая болезни Вегенера, с гранулематозным поражением носа и околоносовых пазух, подтвержденным гистологическим исследованием.

Ключевые слова: Болезнь Вегенер, нефрит, гранулематоз, гранулема носа, васкулит.

CHARACTERISTICS OF DAMAGE TO ENT-ORGANS IN WEGENER'S GRANULOMATOSIS

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ANNOTATION

Wegener's disease, necrotizing granuloma of the upper respiratory tract with nephritis) was first described by the German pathologist F. Wegener in 1936, 1939 [1-5]. A disease from the group of systemic vasculitis, which is characterized by granulomatous lesions, primarily of the upper respiratory tract, lungs and kidneys. Gradually, the disease passes into the second stage, when signs of a generalization of the process join: damage to internal organs (lungs, liver, spleen, kidneys), constant fever, weight loss. The third stage, terminal, is characterized by signs of pulmonary heart and renal failure. In addition to the classical form, Wegener's granulomatosis can proceed according to the "lightning" or "headless" type. The diagnosis is made on the basis of a typical clinical picture. Of the laboratory parameters, a high ESR (40-60 mm/h), normochromic anemia are typical, rheumatoid factor is detected in 50% of patients. Rhinoscopy is important. Histological examination of the biopsy of the nasal mucosa usually reveals small vessel vasculitis, but the most characteristic diagnostic sign is granulomas. Granuloma of the nose (face) malignant (median, fatal) is characterized by damage to the nose, soft tissues of the face, oral cavity, hard palate with the development of extensive ulcerative necrotic defects. The internal organs are usually not affected. During the period from 2020 to 2021, we observed 3 cases of Wegener's disease, with granulomatous lesions of the nose and paranasal sinuses, confirmed by histological examination.

Keywords: Wegener's disease, nephritis, granulomatosis, nasal granuloma, vasculitis.

Introduction. Wegener's disease, necrotizing granuloma of the upper respiratory tract with nephritis) was first described by the German pathologist F. Wegener in 1936, 1939 [1-5]. A disease from the group of systemic vasculitis, which is characterized by granulomatous lesions, primarily of the upper respiratory tract, lungs and kidneys. Some authors distinguish the so-called nasal granulomatosis (malignant, median) as one of the forms of Wegener, but most consider it as an independent disease. In pathogenesis, the main importance is attached to immunological disorders, in particular, deposits of immune complexes in the wall of blood vessels and disorders of cellular immunity. The morphological picture is characterized by necrotic vasculitis of arteries of medium and small caliber, the formation of polymorphic cell granulomas containing giant cells [6].

Wegener's disease manifests itself between the ages of 3 months and 75 years, children, men and women are equally often ill. The average age of patients is 38-40 years [7-10].

Wegener is characterized by the classic triad of symptoms: damage to the upper respiratory tract, lungs and kidneys. In typical cases, the disease begins with damage to the upper respiratory tract (rhinitis, sinusitis). Ulcerative-necrotic rhinitis develops with characteristic complaints of a runny nose with purulent or bloody discharge, dry crusts in the nose. Due to the destruction of the tissues of the nose, its deformation is possible. Ulcerative-necrotic changes are also observed with damage to the paranasal sinuses. In a number of patients, necrotic changes in the mucous membranes of the pharynx, larynx, and ear are noted [11].

Gradually, the disease passes into the second stage, when signs of a generalization of the process join: damage to internal organs (lungs, liver, spleen, kidneys), constant fever, weight loss. The third stage, terminal, is characterized by signs of pulmonary heart and renal failure. In addition to the classical form, Wegener's granulomatosis can proceed according to the "lightning" or "headless" type.

With the "headless" type, the disease is limited only to damage to the lungs and respiratory tract, there is no damage to the kidneys. Naturally, in such cases, the diagnosis of the disease is difficult. A definitive diagnosis is only possible with histological confirmation. The prognosis for life with limited lesions is better than with diffuse spread of necrotizing granulomas [12].

The aim of the work was to study the features of the lesion of the ENT organs in Wegener's granulomatosis.

Material and methods.

The study included 120 patients with Wegener's granulomatosis who applied to the Department of Otorhinolaryngology of the multidisciplinary clinic of the Tashkent Medical Academy.

To examine patients with this pathology, we use clinical (history taking, subjective and objective research methods), clinical and instrumental (radiography and MSCT of PNSs) and laboratory (general blood, bacteriological) research methods.

Results and discussion.

The diagnosis is made on the basis of a typical clinical picture. Of the laboratory parameters, a high ESR (40-60 mm/h), normochromic anemia are typical, rheumatoid factor is detected in 50% of patients. Rhinoscopy is important. Histological examination of the biopsy of the nasal mucosa usually reveals small vessel vasculitis, but the most characteristic diagnostic sign is granulomas. Granuloma of the nose (face) malignant (median, fatal) is characterized by damage to the nose, soft tissues of the face, oral cavity, hard palate with the development of extensive ulcerative necrotic defects. The internal organs are usually not affected. During the period from 2020 to 2021, we observed 3 cases of Wegener's disease, with granulomatous lesions of the nose and paranasal sinuses, confirmed by histological examination.

Among them: men-1; women-2. The mean age of the patients was 37 years. The first sign of the disease in all patients was a lesion of the nose and paranasal sinuses with spread to the orbit. In 1 patient, the process was limited to localization in the nose and maxillary sinuses, which allowed us to regard the disease as a median granuloma; 2 patients later developed a generalization of the process with damage to internal organs (lungs, kidneys, joints).

All patients underwent immunosuppressive therapy in combination with corticosteroids. As a result of the therapy, 2 patients were discharged from the hospital with improvement; ended with a lethality of -1. The study of long-term results of treatment showed, despite the ongoing therapy, life expectancy in 1 patient was 1.5 years, in the second - 3 months. We present the observation of Patient Sh, born in 1979. was on stationary treatment in the ENT department of the OMC from 09.12.20 to 31.12.20, again from 21.01.21 to 26.02.21 with a diagnosis of Wegener's disease with necrotic lesions of the upper jaw and hard palate, orbit. Generalized form

of the 3rd degree, sepsis. Condition after bilateral maxillary sinusectomy and orbitotomy). secondary anemia. Secondary cardiomyopathy on the background of intoxication. Heart failure 1 st. Secondary arrosive bleeding. Encephalopathy. Hypertension-hydrocephalic syndrome.

Upon receipt of a complaint of nasal congestion, the presence of crusts in the nasal cavity, fever up to 39 C for 1.5 months, weakness, lack of appetite. Ill since October 2020, was on outpatient treatment with an ophthalmologist for 2 weeks for conjunctivitis and dacryocystitis, during this period he noted an increase in temperature up to 40C. After examination by a general practitioner, he was sent to the infectious diseases hospital, where from 9.11.20. On November 20, 2020, the patient was hospitalized with a diagnosis of fever of unknown origin. Upon discharge from the hospital, it is recommended: an examination by a phthisiatrician to exclude extrapulmonary tuberculosis, an ENT doctor, an oncologist.

On December 9, 2020, the patient was admitted to the ENT department with a diagnosis of ozena. Fever of unknown origin.

Objectively: General condition of moderate severity. Consciousness is clear, adequate. The skin is pale. Peripheral lymphatic vessels are not palpated. Fever in febrile figures. In the lungs, vesicular breathing, heart sounds of weakened sonority, the rhythm is correct, the pulse is 70 beats. v.min, AD- 100/60 mm. rt st. Nose - The external nose has a regular shape. At the base of the right wing of the nose there is an infiltrate, painful on palpation. At rhinoscopy: the nasal septum in the bone-cartilaginous section is destroyed, there is a defect in the hard palate with a size of 3.0 * 3.2. In the nasal passages, brown crusts with an unpleasant odor, hyposmia. Oropharynx - mucous membrane of normal color, palatine tonsils behind the arches. Submandibular lymph nodes are not palpable. Indirect laryngoscopy - the entrance to the larynx is free, the true vocal folds are gray. During phonation, they close completely. Breathing is free. AD/AS - external auditory canals are free, tympanic membranes are gray, identification borders are clear. There is no pathological discharge. AD/AS-6/6m.

On the survey R-gram of the chest organs in the direct projection from 12/09/20, the x-ray picture is without focal and infiltrative shadows. On the R-gram of the paranasal sinuses dated 10.12.20. bilateral darkening of the maxillary sinuses is determined. CT from 24.12.20. chest organs: no pathology; CT scan of the brain from 01/24/2021. CT features: internal hydrocephalus DEP. Etmoiditis. Frontit.

Ultrasound dated December 27, 2020. abdominal organs: Moderate hepatosplenomegaly. Signs of acute pancreatitis.

Ultrasound of the abdominal organs from 12/23/20. conclusion: reactive changes in the kidneys, splenomegaly. Nephroptosis on both sides.

Ultrasound of the genitourinary system from 21.12.20. conclusion: splenomegaly, pyelectasia of the right kidney, transient form. Nephroptosis on both sides. Ultrasound of the thyroid gland from 22.12.20. conclusion: signs of hypothyroidism of the thyroid gland with moderate diffuse changes.

Echocardiography dated 12/23/20: moderate dilatation of the aorta with slight aortic regurgitation. Mitral valve prolapse of the 1st degree with regurgitation of the 1st degree. Abnormal trabecula in the left ventricle.

Urinalysis dated 12/11/20. color - yellow, transparent - turbidity, specific weight - 1012, reaction - sour, protein - neg., sugar - neg., l - 4-5-6, squamous epithelium - 0.1. urinalysis according to Nechiporenko dated 10.12.04. l - 2.0, er. - no, chi-lindry - no.

Urinalysis dated 18.02.21. color - yellow, proz. - sl.mut, specific weight - cloudy, reaction - sour, protein - neg., sugar - neg.

Blood test from 9.12.20. Hb - 126, er - 4.2, l - 4.0, ESR - 32, L -12, S-81, M-5, P-1, E-1; dated 01/24/20: Nv-108 g/l, l-4.2*10/9, e-3.8*10/12 g/l, ESR-48 mm/h, l-22, s-71, m-5, p-1, e-1, dated 18.02.05. HB - 52 g / l. Er - 2.4 g / l, ESR - 10, C.P - 0.65, B - 1, young - 1, hypochrom - 3, anis. - 3, goodbye. – 3. Evaluation of myelogram dated 27.12.20. Normacytes of all degrees of maturity, megakaryocytic sprout without changes, cell maturation is not disturbed. Blood for sterility from 12/15/20. - no growth, from 24.01.05: staphylococcus epidermis was isolated. OAM (11.12.20). Oud. Weight. 1012, acidic Z - 4-5-6. squamous epithelium 0.1. Biochemical. blood test dated 12/15/20: ALT -69. AST - 61. bilirubin total. 16.4-4.1. alkaline phosphatase 109 units/l. GGTP - 65. cholesterol 3.2, triglycerides 0.71. Total protein - 6, thymol. sample - 4.0 units / mut, urea - 3.4 mmol / l. Creatinine 40 μmol/l, glucose 4.6. LDH - 520, CRP - neg. 12/21/20 atypical cells were not found. Blood test for cytomegovirus dated 04.02.05. negative; from 03.02.05 JqG is negative for chlamydia. Throat swab for flora and sensitivity and antibiotic and fungus. Staphylococcus aureus isolated. 23.02.20 LE -

cells are not found. Microreaction dated 11.02.21; 01/02/21 - negative. Wright-Heddelson reaction from 02/18/21. negative. Antibodies to HIV 1-2 were not detected. CSF analysis from 02/11/21: color - colorless transparent. protein - 0.066 g / l r / i Pandi neg, sugar - 2.3 mmol / l, cytosis - 2/3 * 106 l.

Histopathological examination dated February 10, 21: conclusion - purulent-necrotic inflammation against the background of chronic pseudoepithelimotosis, epithelial hyperplasia with the formation of polyps. Pathological examination dated February 18, 21. productive-necrotic inflammation, exclude specific inflammation, in particular rhinoscleroma and immunodeficiency. Myelogram evaluation from 27.12.20. conclusion: In the smear, normocytes of all degrees of maturity, megakaryocytic sprout without significant changes. Cell maturation is not disturbed.

12/14/20 phthisiatrician. Asset data. Tuberculosis of the lungs is not. 12/22/20 hematologist. Diagnosis: Fever of unknown origin, mild splenomegaly. secondary hypohypochromic anemia stage I; dated 11.02.21. conclusion: secondary hypochromic anemia against the background of osteomyelitis of the nasal bones. 12/29/20 OC AIDS: prolonged fever of unknown origin. Antibodies to HIV were not detected. Oral and maxillofacial surgeon from 04.02.19. a bilateral maxillary sinusectomy was performed. During the operation, thickening of the mucous membrane of the maxillary sinuses, necrosis of bone tissue. Consultation of a neurologist on 01/24/21: Diagnosis: dyscirculatory encephalopathy, hypertension-hydrocephalic syndrome, to differentiate from neuro HIV and Noma of autoimmune genesis. Sepsis. Oku-list dated 24.01.21. -10.02.21, 13.02.21 phlegmon of the orbit. Rheumatologist from 01.02.21 Wegener's syndrome. Rhinitis, osteomyelitis of the bones of the facial skeleton. Council on February 16, 21; from 22.02.21: Wegener's granulomatosis, generalized form 3 st. sepsis with necrotic lesions of the upper jaw and hard palate, the lower wall of the orbit. Secondary septic anemia. Secondary cardiomyopathy against the background of heart failure 1st stage. Ozen.

Treatment: cytostatics, corticosteroids, antibiotic therapy, infusion therapy, vitamins. Treatment without effect. On February 25, 21, nosebleed started, nasal tamponade was performed. Bleeding stopped. 26.02.21 the patient's condition progressively worsened, respiratory and cardiovascular activity stopped. Despite resuscitation measures, biological death was declared.

Conclusion.

A feature of this observation is that the manifestation of the disease was a lesion of the nose and paranasal sinuses, which for a long time was the only symptom of the disease, which made it difficult to diagnose the true lesion. The generalization of the process manifested itself after 2 months. According to the literature, Wegener's disease occurs 1:40,000,000. Despite the fact that the disease is rare, an otorhinolaryngologist needs to know the clinical picture and the course of the disease, since it is difficult to diagnose the disease with the primary local development of the granulomatous process in the nose and paranasal sinuses. Treatment of this category of patients should be under the supervision of a rheumatologist.

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