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WEGENER'S RANULOMATOSIS: CONDITIONS OF THE MUCOUS MEMBRANE ORAL CAVITY AND SKIN (OVEREVIEW)

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ABOUT ARTICLE

Key words: Wegener's granulomatosis, arteriole, vasculitis.

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Abstract: Wegener's granulomatosis (WG) is a rare immunologically mediated multisystem disease of unknown etiology, the morphological basis of which is giant cell granulomatous-necrotizing vasculitis of small arteries, arterioles and venules with combined damage to several organs (mainly the upper respiratory tract, respiratory system, vision, hearing and kidneys). The prevalence of hepatitis B in the population is 25–60 per 1 million, the incidence is 3–12 per 1 million people; in Europe, the prevalence of hepatitis B is five cases per 100 thousand population, and in Northern Europe this figure is even higher.

VEGENER GRANULOMATOZI: OG‘IZ BO‘SHLIG‘I VA TERI QAVATI XOLATI (ADABIYOTLAR SHARHI)

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MAQOLA HAQIDA

Kalit so‘zlar: Vegener granulomatozi, arteriola, vaskulit.

Annotatsiya: Vegener granulomatozi (VG) – noma’lum etiologiyali kam uchraydigan immunologik vositachilik qiluvchi multitizimli kasallik bo'lib, uning morfologik asosi kichik arteriyalar, arteriolalar va venulalarning gigant hujayrali granulomatoz-nekrotik vaskulitlari, bir nechta organlarning (asosan, yuqori ko'rish yo'llari, ko'rish organlarining shikastlanishi, eshitish a’zosi va buyraklar). Aholida VGning tarqalishi 1 million kishiga 25-60, kasallanish 1 million kishiga 3-12; Evropada gepatit B ning tarqalishi 100 ming aholiga beshta, Shimoliy Evropada esa bu ko'rsatkich yanada yuqori.

ГРАНУЛЕМАТОЗ ВЕГЕНЕРА: СОСТОЯНИЕ СЛИЗИСТОЙ ОБОЛОЧКИ ПОЛОСТИ РТА И КОЖИ (ОБЗОРНАЯ ЛИТЕРАТУРЫ)

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О СТАТЬЕ

Ключевые слова: Гранулематоз Вегенера, артериолы, васкулиты.

Аннотация: Гранулематоз Вегенера (ГВ) — редкое иммунологически опосредованное мультисистемное заболевание неясной этиологии, морфологической основой которого является гигантоклеточный гранулематозно-некротический васкулит мелких артерий, артериол и венул с сочетанным поражением нескольких органов (преимущественно верхних дыхательных путей, зрение, органа слуха и почки). Распространенность ГВ в популяции составляет 25–60 на 1 млн, заболеваемость – 3–12 на 1 млн человек; в Европе распространенность ГВ составляет пять случаев на 100 тыс. населения, а в

Северной Европе этот показатель еще выше.

INTRODUCTION

WG (syn.: malignant granuloma, non-infectious necrotic granulomatosis) was first described in 1931 by H. Klinger. Later, F. Wegener (1936, 1939) identified the disease as an independent syndrome with a characteristic triad of signs:

1) systemic necrotizing vasculitis with damage to small-caliber arteries and the venous bed;

2) glomerulonephritis;

3) necrotizing granulomatous vasculitis of the upper respiratory tract (URT) [1].

Currently, the disease appears as an autoimmune granulomatous inflammation of the walls of blood vessels, involving the upper respiratory tract, lungs, eyes, kidneys, skin and other organs. Refers to systemic antineutrophil cytoplasmic antibody-associated necrotizing vasculitis (A) [2].

THE MAIN RESULTS AND FINDINGS

The disease can begin at any age (on average about 40 years), somewhat more often in men, but children rarely get sick. About 15% of patients are under 19 years of age [3].

The etiology of hepatitis B is unknown; chronic focal infection (nasopharyngeal) may play a role. The hyperreactivity of the humoral immunity is important: an increase in serum and secretory immunoglobulins - IgA, IgG and IgE, there are circulating immune complexes, IgG autoantibodies.

The disease is associated with the presence of histocompatibility antigens HLA B7, B8 and DR2, which indicates a certain genetic predisposition. Most patients have antibodies to the cytoplasm of neutrophils, mainly to protease-3 [4].

HV develops gradually: damage to the upper respiratory tract occurs in 92% and is manifested by rhinitis with ulcerative-necrotic changes in the mucous membrane of the paranasal sinuses, larynx, trachea; may be purulent otitis media.

Lung damage is observed in 85–90% of patients and is manifested by cough, shortness of breath, hemoptysis and chest pain. In 1/3 of patients, radiological signs may not be accompanied by clinical manifestations of pulmonary pathology [5-10].

Eye damage, which is observed in 52% of cases, manifests itself in the form of conjunctivitis, dacryocystitis, episcleritis, scleritis, granulomatous sclerouveitis, iridocyclitis, retrobulbar granuloma and exophthalmos.

Heart damage is observed in 8% of cases and leads to pericarditis, coronary vasculitis, myocardial infarction, damage to the mitral and aortic valves, and atrioventricular block.

Damage to the nervous system is observed in 23% of patients and includes neuropathies of the cranial nerves, multiple mononeuropathy, and occasionally cerebral vasculitis and cerebral granulomas.

Kidney damage occurs in 77% of patients and predominates in the clinical picture. It may be limited to mild glomerulonephritis with proteinuria, hematuria and red blood cell casts, but with renal failure it rapidly progresses.

During exacerbations, nonspecific symptoms appear - malaise, weakness, arthralgia, decreased appetite, weight loss, fever [11-13].

Skin lesions are observed in 46% of patients with hepatitis B and are represented by papules, vesicles, and palpable purpura. However, typical manifestations are considered to be nodules and ulcers caused by necrotizing angiitis of the dermal vessels with their thrombosis and necrosis [14-16].

Damage to the oral mucosa occurs in almost all patients and manifests itself as granulomatous growths in the gums, palate, arches, and tonsils. They have a lumpy surface, a stagnant red color, a dense consistency and quickly disintegrate with the formation of ulcers of varying depths. In a number of patients, the process is accompanied by an enlargement of the submandibular lymph nodes. Necrosis of periodontal tissues, perforation of the hard palate, and destruction of soft tissues and bones of the middle third of the face may occur. In this case, when examining the patient, a foul odor is felt [6].

There are two forms of HS – localized and generalized. The first begins with damage to the upper respiratory tract or eyes. Less commonly, the mucous membrane of the mouth and pharynx is primarily affected. In the generalized form, the process begins with damage to the tracheobronchial tree and lungs and is clinically manifested by fever of varying severity, polymorphic rashes, and a cough with purulent-bloody sputum. Then symptoms of damage to other organs appear. Possible arthritis, arthralgia and myalgia, anemia, neutrophilic leukocytosis, accelerated ESR. Chondritis of the auricles, etc. may develop.

The prognosis of the disease is unfavorable. Without timely treatment, death can occur within 6–12 months. Death occurs more often from renal or cardiovascular failure [4].

Diagnostics

25% of patients in the initial stage have no signs of kidney or lung damage, and only in 50% of patients with hepatitis B is diagnosed in the first 3–6 months from the onset of the disease, and in 7% this disease is not diagnosed even within 5– 16 years from the appearance of the first clinical symptoms.

Typical laboratory findings in hepatitis B are: a significant increase in ESR, anemia, leukocytosis, hypergammaglobulinemia (mainly due to IgA), the appearance of rheumatoid factor,

urinary syndrome characteristic of glomerulonephritis, antibodies to protease-3 are found in 90% of patients with lesions respiratory tract and kidneys and only in 70% of patients without kidney damage (T.V. Beketova, 1995).

The histological diagnosis of hepatitis B is characterized by the detection of necrotizing vasculitis in the biopsy specimen, accompanied by granulomatous inflammation [8,9].

The differential diagnosis for a dermatologist is made with median facial granuloma and lymphomatoid granulomatosis.

Median granuloma of the face affects only the upper respiratory tract, including the paranasal sinuses, and is accompanied by extensive destruction of soft tissues and ulceration of the facial skin, which is not typical for hepatitis. Inflammation and necrosis can involve vessels, but vasculitis is almost never primary.

Lymphomatoid granulomatosis is a disease from the group of angiocentric lymphomas. The disease affects the lungs, skin, central nervous system and kidneys, where there is infiltration of the walls of blood vessels and surrounding tissues with atypical lymphocytes and plasma cells. In contrast to hepatitis B, in lymphomatoid granulomatosis there is no vasculitis as such, but the formation of granulomas is noted. In more than 1/2 of patients, the disease becomes frankly malignant. In the differential diagnosis of hepatitis B, determining the titer of antibodies to protease-3 is of great importance [17-21].

Differential diagnosis must also be made with other vasculitis, especially with Churg-Strauss syndrome, as well as with Goodpasture syndrome, tumors of the upper respiratory tract and lungs, mucocutaneous leishmaniasis, scleroma, and other infectious and non-infectious granulomatosis.

Treatment

Since the disease is of autoimmune origin, the drugs of choice are immunosuppressants: cytostatics in combination with glucocorticoids. Of the cytostatics, cyclophosphamide at a dose of 2 mg/kg per day has proven itself to be effective. Glucocorticoids are prescribed in a dose equivalent to prednisolone 1 mg/kg per day. Pulse therapy with cyclophosphamide at a dose of 1000 mg showed good results. There are reports of a positive effect of treatment with human donor polyvalent Ig. Certain hopes are placed on anti-cytokine therapy [10].

CONCLUSION

Chronic granulomatous inflammation of the oral mucosa is a local form of hepatitis B.

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