www.journalofresearch.us
info@journalofresearch.us

HEALTH SCIENCE

Manuscript info:
Received September 17, 2018., Accepted October 16, 2018., Published October 30, 2018.

FEATURES OF THE COURSE OF WEGENER'S GRANULOMATOSIS IN COMBINATION WITH COMORBID PATHOLOGY

Rizamukhamedova Mashhura Zokirovna,
Djuraeva Elnora Rustamovna, Berdieva Dilfuza Umurzakovna,
Tashpulatova Maktuba Mukhamedalievna,
Tashkent Medical Academy

http://dx.doi.org/10.26739/2573-5616-2018-10-2-9

Abstract: Wegener's granulomatosis (WG) - a rare systemic disease of unknown etiology. Histopathological characterized by granulomatous necrotizing systemic vasculitis, most often involving the upper and lower airways with further lung damage. WG debut in most cases differs no specificity of clinical picture and the complexity of timely diagnosis, which leads to a delay of treatment and poor prognosis. Presented 1 case of WG caused considerable difficulties in diagnosis at the early stage of the disease.

Keywords: systemicvasculitis, Wegener's granulematosis

Recommended citation: Rizamukhamedova Mashhura Zokirovna, Djuraeva Elnora Rustamovna, Berdieva Dilfuza Umurzakovna, Tashpulatova Maktuba Mukhamedalievna. FEATURES OF THE COURSE OF WEGENER'S GRANULOMATOSIS IN COMBINATION WITH COMORBID PATHOLOGY. 9-10 American Journal of Research P. 86-90 (2018).

Wegener's granulomatosis (WG) is a systemic vasculitis of the arteries and veins of the middle caliber, characterized by the development of necrotizing granulomatous inflammation with a predominant lesion of the upper respiratory tract, lungs and kidneys [1]. WG was first described in 1931 by H. Klinger (1936) and F. Wegener (1939)[5]. Both men and women are sick often in the age of about 40 years. [4, 9].

The etiology of the disease is unknown. It is assumed that the onset of the disease is related to the effect of infectious agent. Some researchers note a link between WG with a previous purulent infection or tuberculosis of the respiratory tract. A more severe course of WG in patients with S. staphylococcus aureus (Staphylococcus aureus) carriage in the nasal cavity is shown, which is characterized by more frequent exacerbations associated with exposure to exotoxins of staphylococcus possessing the properties of super antigens. The potential etiological role of colloidal silicon compounds is also discussed. [5].

Pathogenesis of WG is associated with the development of widespread inflammation of small vessels and the simultaneous formation of perivascular and extravasal granulomas of macrophage type with Langhans cells in the affected organs and tissues. The serologic marker of the disease is antibodies to the cytoplasm of neutrophils that bind to antigens expressed by neutrophils (proteinase-3, myeloperoxidase, etc.), and can cause degranulation of these cells with the release of proteolytic enzymes. Other pathogenetic mechanisms are discussed: immune complex lesion of the vascular wall, lymphocytic cytotoxic reactions, etc. [3].

The clinical picture of WG [1] is characterized by multiple organ dysfunction. With WG, more than 90% of the cases show signs of upper respiratory tract, lung and middle ear injuries. Sinusitis is observed in 50% of patients, in 36% - rhinitis, in 10% pleurisy, in 25% media otitis. Upper respiratory tract and ear are affected in 92%, lungs - in 85% of patients. [7].

1. Glomerulonephritis develops in 75-80% of patients, but it is rarely the first manifestation of WG.

2. The defeat of the musculoskeletal system is detected in about 70% of patients.

3. In 52% of cases with WG, eye damage is observed - from mild conjunctivitis to dacryocystitis, iridocyclitis, granulomas of retrobulbar fiber and exophthalmos. [8].

4. Skin lesion is noted in 46% of patients and includes papules, vesicles, palpable purpura, ulcers, subcutaneous nodules. Vasculitis, granulomas or both are revealed in biopsy.

5. The defeat of the heart is observed in 8% of cases and leads to pericarditis, coronary vasculitis, myocardial infarction, damage of mitral and aortic valves, and atrioventricular blockade.

6. The defeat of the nervous system is noted in 23% of patients and includes polyneuropathy, cerebral vasculitis and cerebral granulomas. [1,6].

"Gold standard" - histological examination of affected tissue (nasal mucosa, lung tissue, skin or kidney, granulation tissue of the orbit of the eye) which obtained by biopsy, with the detection of fibrinoid necrosis and inflammation of the vessel wall in combination with perivascular and extravasal granulomas [2,11]. Characteristic changes in laboratory indicators: normochromic anemia, thrombocytosis, neutrophilic leukocytosis, acceleration of ESR, increase of the level of CRP. ANCA is identified in 50-70% of patients, which considered as a factor of unfavorable prognosis of the disease. RF is detected approximately in every second patient, which is a nonspecific marker of activity of WG. The level of Ig in serum, as a rule, within the limits of the norm, an increase of the content of the CIC, hypocomplexemia, the appearance of AT to the glomerulus membrane is not characteristic.

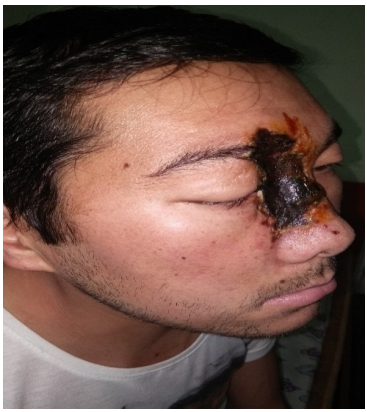
According to the literature data, the mortality rate of WG is still high.

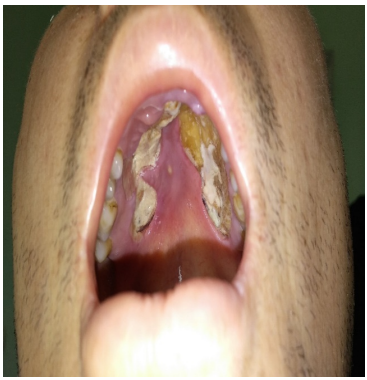
The most common causes of death are intercurrent infections, respiratory and renal failure, cardiovascular disorders, malignant neoplasms (bladder cancer). This causes the need for timely diagnosis of the disease with the subsequent early appointment of aggressive therapy before the development of irreversible damage to vital organs. WG must be suspected in all patients with fever, weight loss and signs of multi-organ damage (upper respiratory tract, lungs, urinary syndrome, vascular purpura, multiple mononeurotic). The five-year survival rate is more than 65% with adequate therapy of WG [7].

We present the clinical case of WG in combination with 1 type of diabetes mellitus, which caused difficulties in diagnosing the disease.

Patient D., male, 27 years old, has been suffering from illness since March 2018, he began to notice pain and swelling in the left side of the face, mucous discharge from the nose after hypothermia. A few days later, there was a sub febrile condition, the visual acuity on the right eye decreased. Since the patient suffered 1 type of diabetes during the 1st year, he turned to endocrinologist and he was hospitalized in the department of endocrinology with a diagnosis of diabetic retinopathy. Monitoring of laboratory parameters in the blood test indicated an increase of ESR up to 72 mm / h, leukocytosis (20*109/L), glucose hemoglobin level-9.2 mmol / L, glycemic hemoglobin-9.4%, in general urine analysis proteinuria, cylindruria, when determining acute phase samples: CRP-24 mg/l. Radiography of the lungs: the roots of the lungs are expanded and compacted, the pulmonary fields are clean. MRI of the brain: pathological changes were not detected. The patient has received hypoglycemic, metabolic drugs and agents that improve peripheral circulation. However, the therapy did not yield any significant results. Ulcerative necrotic eruptions with merging character in the region of the right half of the back of the nose, the right eyelid and on the mucosa of the hard palate appeared during the stay in the hospital. There were mucopurulent discharge from the nose with the formation of bloody crusts, which made it difficult for the nasal breathing, worried about aching pains in the region of both knee joints that were not accompanied with swelling. Otolaryngologist and ophthalmologist examined the patient, eventually, consultation of the maxillofacial surgeon was recommended. The diagnose WG was put to patient and recommended treatment in the rheumatology department. At admission, the condition of the patient was medium severity, the consciousness is clear. Speech hoarseness is noted. Body temperature was 37.40C. Saddle-shaped deformation of the nose was indicated. The skin was pale, in the region of the right half of the back of the nose, the right eyelid, there are ulcerative necrotic eruptions with indistinct boundaries, in the oral

cavity on hard palate-ulceration, shaped like oval with pronounced red borders and grayish yellow coating in the center.





Moderately enlarged submandibular lymph nodes were palpable. Edema of the lower leg on both sides. Palpation of knee joints is painful. In the lungs, breathing is weakened vesicular breathing, wheezing is not heard. BR-20 in min. Heart area without features. Heart sounds are rhythmic, muffled. HRR-114 in min, AP-110/70 mmHg. The abdomen is painless in palpation, the liver + 1cm from under the right costal arch. The spleen is not enlarged. Stool and diuresis are not violated. A clinical diagnosis: Systemic vasculitis. Wegener's granulomatosis, generalized form, acute course, high activity, with damaging of upper respiratory tract (destruction of the cartilaginous and bony tissue of the nasal septum, hard palate), eyes, kidneys (secondary glomerulonephritis with nephrotic syndrome), heart. Concomitant diagnosis: 1 type of diabetes mellitus, decompensated stage.

In consideration of the high clinical and laboratory activity of the disease, a combined pulse therapy with 500 mg of solumedrol and 1000 mg of cyclophosphamide №2 was performed. Methylprednisolone was administered intravenously in a dose of 40 mg per day, cyclophosphamide intravenously dripped in a dose of 200 mg per week, azathioprine intravenously in a dose of 100 mg per day. The tolerability of the therapy was satisfactory. Positive dynamics was noted. Completely docked signs of rhinitis, the boundaries of necrotic foci became clearly delineated. In the future, the patient was recommended a programmed pulse therapy.

Thus, this clinical case demonstrates the multifaceted manifestations of WG, the tendency of the disease to develop a generalized form with the defeat of vital organs, which significantly

worsens the prognosis, as well as the complexity of diagnosing the disease in the presence of a comorbid pathology, in particular diabetes mellitus. Timely active treatment of WG has a decisive influence on the course of the disease and can prevent disability of the patient.

References:

1. Zubkova L.L., Baldanova I.R., Borhonova I.V., ErdyneevaE.B. Wegener's granulomatosis (case from practice) // Bulletin of the Buryat State University - 2009/12. - 99-101

2. Mesova A.M., Sagymbaeva G.K., Kazhiekberova A.T., Kozachenko A.B., AkhmetovaZh.B. Granulomatosis of Wegener (clinical case) // Bulletin of KazNMU. - 2016. - №1. - P.71-73.

3. Rheumatology: clinical recommendations / ed. acad. RAMS E.L. Nasonov. - 2 nd ed., Pererab. and additional. - Moscow: GEOTAR-Media.-2010. -752 sec.

4. Sigitova O.N., Bikmoukhammetova E.I., Bogdanova S.R. Granulomatosis of the wegener (clinical case from practice) // Bulletin of modern clinical medicine. - 2010. - Том 3. № 3. - C.73-76

5. Allen S.D., Harvey C.J. Imaging of Wegener's granulomatosis // The British Journal of Radiology. - Vol. 80. - 2007. - P. 757-765.

6. Guillevin L. Mahr A. Wegener's granulomatosis // Orphanet encyclopedia. - 2004. - P. 1-5.

7. Khan A.R., Chapman, P.T., Stamp, L.K., Wells J.E. and O'Donnell J.L. Wegener's granulomatosis: treatment and survival characteristics in a high-prevalence southern hemisphere region // Internal Medicine Journal. - 2012 - Vol. 42. - P. 23-26.

8. Kubal A.A., Perez V.L. Ocular manifestations of ANCA-associated vasculitis // Rheum. Dis. Clin. North. Am. - 2010. - Vol. 36, № 3. - P. 573-586.

9. Mahr A.D., Neogi T., Merkel P.A. Epidemiology of Wegener's granulomatosis: Lessons from descriptive studies and analyses of genetic and environmental studies and analyses of environmental risk determinants //Clin.Exp.Rheumatology. - 2006; - 24 (Suppl. 41): P 82-91.

10. Pagnoux C., Wolter N.E. Vasculitis of the upper airways // Swiss. Med. Wkly. - 2012. -Vol. 142. - P. 135-141.