

SYSTEMIC VASCULITIS: GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS)

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Abstract

Granulomatosis with polyangiitis (GPA) is a disease, based on autoimmune granulomatous inflammation of vascular wall (vasculitis), involving small and medium blood vessels – arteries, arterioles, venules and capillaries of upper respiratory tract, eyes, kidneys, lungs and other organs. GPA belongs to heterogeneous group of systemic vasculitis.

The care for patients with GPA is provided by rheumatologists. At the same time, clinical picture of GPA includes the symptoms attributed to the lesions of upper airways, trachea, bronchi and lung parenchyma, which make the patients to refer to HEENT physicians and pulmonologists. Thus, the problem of the diagnosing of GPA has the multidisciplinary background.

This review defines the place of GPA in current nomenclature of systemic vasculitis, adapted at 2nd International consensus conference (Chapel-Hill) in 2012, referring the issues of epidemiology, pathogenesis, morphology, clinical course, diagnosis and treatment.

The authors highlighted the pathomorphology of GPA, possible etiology and structure of granuloma and infiltrates, determining clinical manifestations, a course and the outcome of the disease.

The article is dedicated to practicing pulmonologists, rheumatologists, physicians and radiologists.

Key words: systemic vasculitis, granulomatosis with polyangiitis, pathogenesis, pathomorphology, clinical symptoms, diagnosis, treatment.

Ukr. Pulmonol. J. 2019;3: 52–57.

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