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## Granulomatosis with polyangiitis–Clinical characteristic and treatment

Granulomatosis with polyangiitis (GPA) is a rare systemic disease characterized by granuloma formation in small and medium sized vessels, inflammatory changes and necrotic tissue formation. Blood analysis reveals the presence of ANCA antibodies against proteinase 3 (PR3-ANCA). The etiopathogenesis of GPA notes the importance of IL-1, IL-12, IL-18 cytokines, TNF-alpha, INF-gamma and PR3-ANCA antibodies. Clinical features include dominant upper respiratory tract symptoms (inflammation of the nasal mucous, sinusitis, middle and inner ear inflammation, laryngitis and inflammation of the trachea). The most dangerous symptoms is subglottic stenosis, which should be treated immediately. Other symptoms include bronchitis and lung diseases (pulmonary nodules leading to cavity formation, pulmonary infiltrates and haemorrhage). Glomerulonephritis may lead to renal failure, Ocular changes (scleritis, episcleritis, iritis, corneal inflammation sometimes leading to its perforation, conjunctivitis, lacrimal duct changes, pseudo tumor in the orbit which may lead to blindness and inflammation of the opthic nerve) Myalgia, arthritis, purpura, subcutaneous skin nodule, ulcers, necrosis of digits, mononeuropathies, polyneuropathies, cranial nerve damage, ischemic stroke, cerebral hemorrhage, endocarditis, pericarditis, abdominal pain, diarrhea and gastrointestinal haemorrhage may occur. The clinical form of GPA present as local symptoms (upper or lower respiratory tract without systemic symptoms), early systemic disease (disease without compromising organ function and death), generalized systemic type (disease with organ dysfunction, creatinine concentration<500 umol/L), severe type (disease with multiorgan failure, creatinine concentration>500 umol/L), resistant type (disease progression despite treatment with glucocorticosteroids and cyclophosphamide). Treatment of local symptoms and early systemic disease includes immunosuppressive drugs such as methotrexate or azathioprine with medium dose of glucocorticosteroids. The generalized systemic type, resistant type and severe type of disease is treated using high doses of glucocorticosteroids and cyclophosphamide administered parenterally. In rapidly progressive glomerulonephritis, plasmapheresis is considered. A new treatment regime using rituximab in doses of 375 mg/m$^2$ once a week for four weeks is suitable.

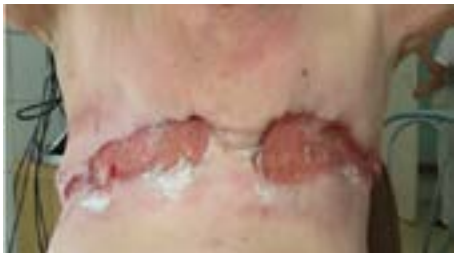

Figure 1: File Patient with GPA.ppt enclosed

### Recent Publications

1. Geetha D, Kallenberg C, Stone J H, et al., (2016) Current therapy of granulomatosis with polyangiitis and microscopic polyangiitis: the role of rituximab. Journal of Nephrology 28:17-27.
2. Kallenberg C G M (2015) Pathogenesis and treatment of ANCA-associated vasculitides. Clinical and Experimental Rheumatology 33(92):S11-S14.
3. Jennette J C, Falk R J, Bacon P A, et al., (2013) 2012 revised international chapel hill consensus conference nomenclature of vasculitides. Arthritis & Rheumatology 65:1-11.

4. Specks U, Merkel P A, Seo P, et al., (2013) Efficacy of remission-induction regiments for ANCA-associated vasculitis. New England Journal of Medicine 369: 417-427.

5. Jennette J C, Falk R C, Thurston D, et al., (2011) Pathogenesis of ANCA vasculitis. Current Opinion in Nephrology and Hypertension 20(3):263-270.

## Biography

Małgorzata Wisłowska is the Head of Department of Rheumatology and Internal Medicine of Central Clinical Hospital MSWiA Warsaw, Poland. She is a specialist in Internal Medicine, Rheumatology, Rehabilitation Medicine, Hypertension and the author of over 200 scientific papers and books. She has participated in numerous scientific meetings and is a promoter of 12 PhD theses. She took training at Guy and St. Thomas' Hospitals in London, Charity Hospital in Berlin, Rheumatology Institutes in Prague and Moscow. In 2003, she started the Department of Internal Medicine and Rheumatology and in 2010 the Clinic of Internal Medicine and Rheumatology CSK MSW. She is a Professor at the Warsaw Medical University. Her research interests include "Internal medicine, rheumatology, rehabilitation medicine and hypertension".

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