Granulomatosis with polyangiitis

Granulomatosis with polyangiitis (formerly **Wegener's granulomatosis**[1]) is a necrotizing vasculitis of small vessels that affects:

- **respiratory system** by the formation of granulomas,
- **kidneys** necrotizing glomerulonephritis.

Clinical picture

General symptoms
: febrile, weight loss, fatigue;

ORL area
: inflammation (up to necrotizing) of the upper respiratory tract with the formation of ulcers, epistaxis, sinusitis, destruction of the nasal cartilages with the formation of the so-called saddle nose, subglottic stenosis of the trachea manifested by stridor with danger of acute asphyxia, chronic carrier of *Staphylococus aureus*; otitis media, conductive hearing disorders;

Lower respiratory tract
: cough, chest pain, hemoptysis from typical necrotizing granulomas in the bronchi;

Kidney
: rapidly progressing glomerulonephritis (ANCA-positive glomerulonephritis) with an acutely developing picture renal insufficiency;

Gastrointestinal tract: diarrhoea, enterorrhagia, abdominal pain, endoscopically demonstrable haemorrhages and ulcerations;

Peripheral nerves
: mononeuritis multiplex;

Eyes
: inflammation of the cornea with the formation of ulcers, danger of blindness;

Locomotor system: arthralgia, myalgia, erosive arthritis.

Diagnostics

Laboratory findings:

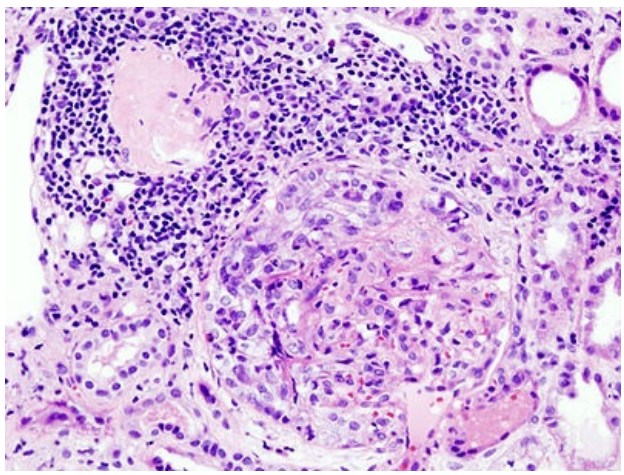
- ↑ PAF, ↑ FW, hypergammaglobulinemia;
- anemia, leukocytosis, thrombocytopenia;
- proteinuria, erythrocyturia
- **c-ANCA** antibodies (positivity recedes as disease activity subsides);
- **rheumatoid factors** are demonstrated in some patients;
- **granulomatous inflammation'** in a biopsy specimen.

X-ray of lungs:

- Butterfly shading or nodal shading.

Renal biopsy

- Focal to focal-segmental glomerulonephritis;
- histologically, sometimes demonstrable vasculitis of small vessels.



Glomerulonephritis in an ANCA positive patient.

Therapy

- Active forms: combined pulse treatment with methylprednisolone + cyclophosphamide as in PAN;
- in case of positivity of ANCA antibodies and kidney involvement or hemoptysis: immediate plasmapheresis.

Prognosis

- Depends on the degree of kidney involvement, 90% of patients survive an average of five years;
- untreated disease has a poor prognosis: up to 70% of patients die;
- has a tendency to "relapses", they are frequent, they appear in up to 50% of patients even several years after diagnosis, often in connection with infection or reduction of corticosteroid doses, *large relapses* are treated with pulsed application of methylprednisone and cyclophosphamide, *minor relapses* by increasing the maintenance dose.

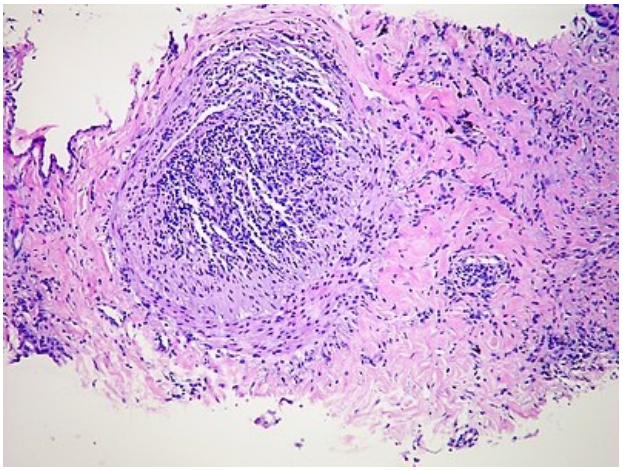
Links

Related Articles

- Autoimmune disease
- Glomerulonephritis
- Rapidly progressive glomerulonephritis
- Systemic vasculitides

References

- KLENER, P, et al. *Vnitřní lékařství.* 3. edition. Praha : Galén, 2006. ISBN 80-7262-430-X.



Arthritis-granulomatous inflammation and necrosis in a patient with Wegener's granulomatosis

References

1.

Missing link to the original WikiScripta article