Granulomatosis with polyangiitis

General information

Formerly known as Wegener's granulomatosis.A systemic necrotizing granulomatous vasculitis involving the respiratory tract (lung → cough/hemoptysis, and/or nasal airways → serosanguinous nasal drainage ± septal perforation→ characteristic "saddle nose deformity") and frequently the kidneys (no reported cases of kidney involvement without respiratory).

Nasal obstruction and crusting are the usual initial findings. Arthralgia (not true arthritis) is present in > 50%.

Neurologic involvement usually consists of cranial nerve dysfunction (usually II, III, IV, & VI; less often V, VII, & VIII; and least commonly IX, X, XI, & XII) and peripheral neuropathies, with diabetes insipidus (occasionally preceding other symptoms by up to 9 months). Focal lesions of the brain and spinal cord occur less frequently.

Differential diagnosis

Differential diagnosis includes:

● "lethal midline granuloma" (may be similar or identical to polymorphic reticulosis) may evolve into lymphoma. May cause fulminant local destruction of the nasal tissue. Differentiation is crucial as this condition is treated by radiation; one should avoid immune suppression (e.g. cyclophosphamide). Probably does not involve true granulomas. Renal and tracheal involvement do not occur

● fungal disease: Sporothrix schenckii & Coccidioides may cause identical syndrome

● other vasculitides: especially eosinophilic granulomatosis with polyangiitis (EGPA)

(asthma and peripheral eosinophilia usually seen), and PAN (granulomas usually lacking)

Evaluation

Biopsy of upper airways consists of removing all crusts, and obtaining as much friable mucosa as possible. This tissue should be fixed in formaldehyde and examined pathologically within 24 hrs (do not freeze). A sample should also be cultured (including fungal and acid-fast cultures). Renal biopsy should not be done when more specific tissue is available from the upper airway.

Elevated serum levels of autoantibodies to proteinase 3 (PR3) antigen are characteristic for GPA, and are detected in 95% of histologically proven cases.

Treatment

Granulomatosis with polyangiitis treatment