Wegener's granulomatosis of the head and neck - our experience
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The aim of our poster is to present several cases of Wegener’s granulomatosis (WG) patients with head and neck manifestations including epistaxis, nasal crusting, prolonged sinus infections with atypical microbiological causes and nasal polyps.

Materials and methods: These WG patients were referred to our clinic in 2016 for evaluation on the account of their complaints. Among the used methods were full ENT examinations, CT scan of neck and mediastinum, fibrolaryngoscopy, serological testing for ANCA antibodies.

Case 1 43- yo male patient presenting with severe recurrent pansinusitis unresponsive to several oral antibiotics over two months. Microbiological culture of sinus discharge showed Chryseobacterium meigeosepticum, an extremely rare cause of sinusitis as it is a common cause for meningitis and pneumonia in children and immunocompromised patients. After a 10-day treatment in accordance with the antibiogram with ciprofloxacin the sinonasal symptoms subsided and normal flora was cultured but the patient developed dyspnea, cough and fever. CT scan showed lung inflammation, PR3-ANCA tested positive and he was started on a CS pulse with significant improvement.

Case 2 A 66- yo patient presenting with severe nasal inflammation, sanguinous crusting and septal perforation. Treated locally to prevent crusting and eliminate bacterial colonisation. Tested positive for C-ANCA and MPO-ANCA. Again, successsfully treated with remission with CS.

Discussion Wegener’s granulomatosis is an idiopathic systemic inflammatory disease typically presenting with vasculitis affecting the upper and lower respiratory tracts and the kidneys. The classical histopathologic features include granulomatous inflammation, necrosis, and/or vasculitis. The initial symptoms typically involve the ENT region. The nose and sinuses are the most frequently affected sites in the head and neck. Symptoms of nasal mucosa inflammation include crusting, pain, epistaxis, ulceration, altered sense of smell, serosanguinous or purulent discharge, and nasal obstruction,

septal perforations and saddle nose deformity; The middle ear is affected in at least one-third to one-half of all patients (2). Clinical features include otalgia, oitis media, and serous, purulent, or bloody discharge. Patients with chronic otitis media may present with conductive hearing loss due to thickened, scarred, or perforated tympanic membranes. Chronic otitis media can also lead to the development of a cholesteatoma. Subglottic stenosis can occur as a result of scar formation in 20 %

About 80% of all patients with WG are ANCA-positive and 80% of those patients produce ANCA with a diffuse cytoplasmic fluorescence pattern (C-ANCA) and antigen specificity for proteinase-3 (PR3-ANCA). About 10% to 20% of ANCA may have a perinuclear fluorescence pattern (P-ANCA) with specificity for myeloperoxidase (MPO-ANCA). The sensitivity of PR3-ANCA in WG is approximately 90% in patients with severe active disease and 55% to 75% in those with milder active disease

The differential diagnosis of ENT-related WG disease includes chronic infections (e.g., tuberculous, fungal, and syphilitic), malignancy (e.g., lymphoma), sarcoidosis, Churg-Strauss syndrome. The differential diagnosis of septal perforations associated with WG includes sarcoidosis, cocaine use, SLE, extranodal nasal lymphoma, lymphomatoid granulomatosis (LYG), and excessive use of intranasal corticosteroids.

Features of disease
Prevalence at onset of disease (%) Prevalence during course of disease (%)
Ear, nose, and throat 70 90
Lung 45 85
Glomerulonephritis 20 70–80
Eye 15 50
Skin 15 45
Musculoskeletal 30 70

References