

Headmirror's ENT in a Nutshell

Eosinophilic Granulomatosis with Polyangiitis (EGPA)

Churg-Strauss Syndrome

Experts: Garret Choby, M.D. & Matthew Koster, M.D.



Presentation (0:33)

- Symptomatology
 - Sinonasal dysfunction with chronic rhinosinusitis with nasal polyps
 - Long standing asthma or adult-onset asthma
 - Serous otitis media with possible sensorineural hearing loss
 - **Prodromal phase**
 - Asthma, nasal polyposis, weight loss, fever, malaise
 - **Eosinophilic phase**
 - Rise or further increase in peripheral eosinophils
 - >10% of leukocytes or > 1500 absolute eosinophil count
 - Deposit in organs leading to worsening lung function or sinonasal disease
 - **Vasculitic phase**
 - Skin nodules (palpable purpura)
 - Alveolar hemorrhage
 - Mononeuritis multiplex
- Differential Diagnosis
 - Aspirin exacerbated respiratory disease
 - CRSwNP and asthma
 - Allergic fungal sinusitis
 - Hypereosinophilic syndrome
 - Allergic bronchopulmonary aspergillosis

Pathophysiology (4:55)

- Genetic predisposition of different polymorphism that increases likelihood for this but requires additional stimulus (environment, infection, drug, inhaled antigen)
- Two pathways
 - Interleukin-5 (IL-5) up-regulation: increases eosinophilic progenitors which mature and then release into blood stream and deposit into the tissues leading to local inflammation and organ damage (sinuses, GI tract, cardiac structures). TH2 pathway.
 - ANCA mediated inflammation in which neutrophils get primed and increase pro-inflammatory mediators that lead to cyclic pathway of neutrophil priming and inflammation (kidneys, lungs, nerves)

Workup (9:14)

- Thorough history and physical exam to evaluate different organ systems that may be related
 - EGPA is a clinical diagnosis
 - Biopsy is confirmatory

- Laboratory studies
 - CBC (eosinophilia) → will trend these
 - >10% of leukocytes
 - ANCA studies (40-60% patients will be ANCA positive)
 - Likely pANCA
 - Negative ANCA with high clinical history does not rule out
 - Renal function
- Imaging
 - Chest Xray
 - CT sinus
 - Echocardiogram and/or cardiac MRI for myocarditis (if indicated)
 - CT abdomen (if chronic diarrhea)
- Additional studies / evaluations
 - Pulmonary function testing with methacholine challenge
 - Nitric oxide testing can demonstrate asthma as well
 - Neurologic evaluation
- **Classification criteria to consider (no specific diagnostic criteria)**
 - **Asthma, eosinophil >10% leuk, mono/polynuropathy, pulmonary opacities, paranasal sinus abnormalities, extravascular eosinophil (bx of vessel)**

Treatment (15:10)

- Five factor score
 - Heart, lung, GI and central nervous system
 - One or more involvement is considered severe disease and requires induction with cyclophosphamide and high dose steroids
 - Score of zero (mild disease) may be treated with moderate dose of prednisone with taper
- Most patients have recurrence of asthma and sinusitis and will respond to steroid
 - Relapses will need disease modifying agents (methotrexate, azathioprine, mycophenolate)
- Biologics
 - Mepolizumab (NUCALA)
 - Inhibitor of IL-5 (great for asthma or sinus disease)
 - Does not work well for patients with vasculitic involvement (ANCA driven)
 - May require cyclophosphamide or rituximab
- Sinus Disease
 - Treat with medical/surgical management like typical CRS patients