

**CME: Managing Individuals with Eosinophilic
Granulomatosis with Polyangiitis (EGPA)**

Module 3: Diagnostic Challenges

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Diagnostic Challenges

EGPA is a rare disease

- In the U.S., rare disease defined as affecting >200,000 people
- ~ 3000 – 4000 people in U.S. have EGPA (10-15 cases per 1 million)
- Not well known
- Symptoms mimic other more common ailments

Symptoms

Symptoms mimic other more common ailments

- Asthma
- Allergies
- Rash



EGPA is Not Well Known

Most patients do not know about EGPA

- Not something you can google

Most clinicians do not know about EGPA

- Extremely rare
- Very limited published articles

Most clinicians not familiar with eosinophils

- Eosinophils > 1200/ml

Groh et al. *Eur J Intern Med* 2015



Work Up

History and Physical Exam

- Thorough history and exam needed
- Unexplainable neuropathy, rash, or respiratory functions should be further investigated
- Eosinophil counts tend to be overlooked



Differential Diagnosis

Broad Initial Differential Diagnosis: infections, drug reactions, malignancies, and many rare diseases

- Serologic testing for toxocariasis and human immunodeficiency virus (HIV)
- Specific IgE and IgG dosages for *Aspergillus* spp.
- Test for *Aspergillus* spp. in sputum and/or bronchoalveolar lavage fluid
- Tryptase and vitamin B12 levels
- Peripheral blood smear (looking for dysplastic eosinophils or blasts)
- Chest computed-tomography (CT) scan

Differential Diagnosis

Other eosinophilic disorders

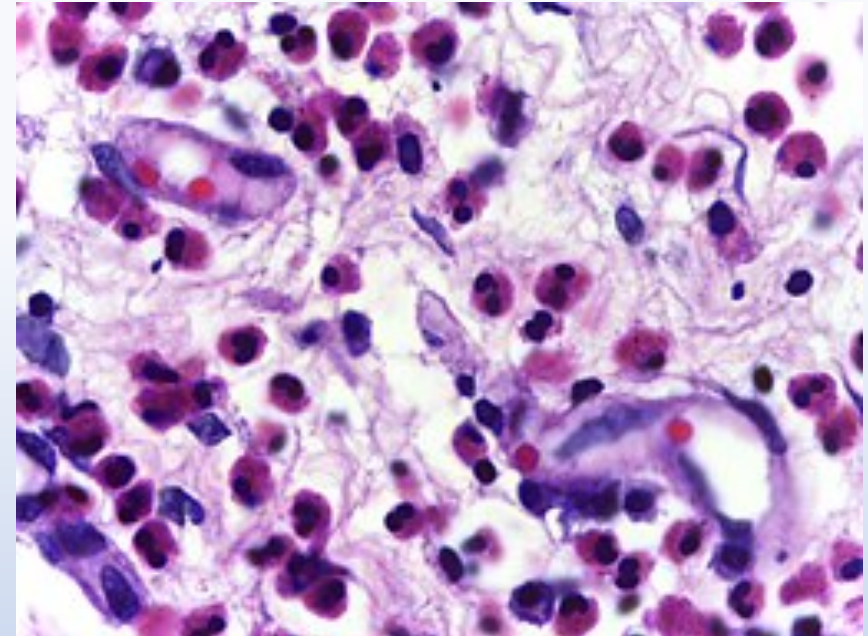
- Eosinophilic asthma
- Eosinophilic pneumonia
- Hypersosinophilic syndromes

Causes of reactive hypereosinophilia

- Drug-induced? Travel/geography?
- *Toxocar?* *Strongyloides stercoralis?* *Aspergillus?*
- Cancer or family history of cancer?

Cutaneous lesions

- Hypergammaglobulinemia
- Gleich's syndrome



Classifications

1990 ACR criteria for Churg-Strauss syndrome

- Asthma
- Eosinophilia (>10% of total WBC)
- Neuropathy
- Pulmonary infiltrates non-fixed
- Paranasal sinus abnormalities
- Extravascular eosinophils

2012 North Carolina Consensus

- Churg-Strauss syndrome renamed EGPA
 - ANCA

Summary

Key takeaways

- Many diagnostic challenges
- New classifications in place
- History and physical exam key to differential diagnosis