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

Module 2: Signs and Symptoms

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Phasic Presentations

People with EGPA often present in three phases

1. Asthma/allergy phase
2. High eosinophil phase
3. Vasculitis phase

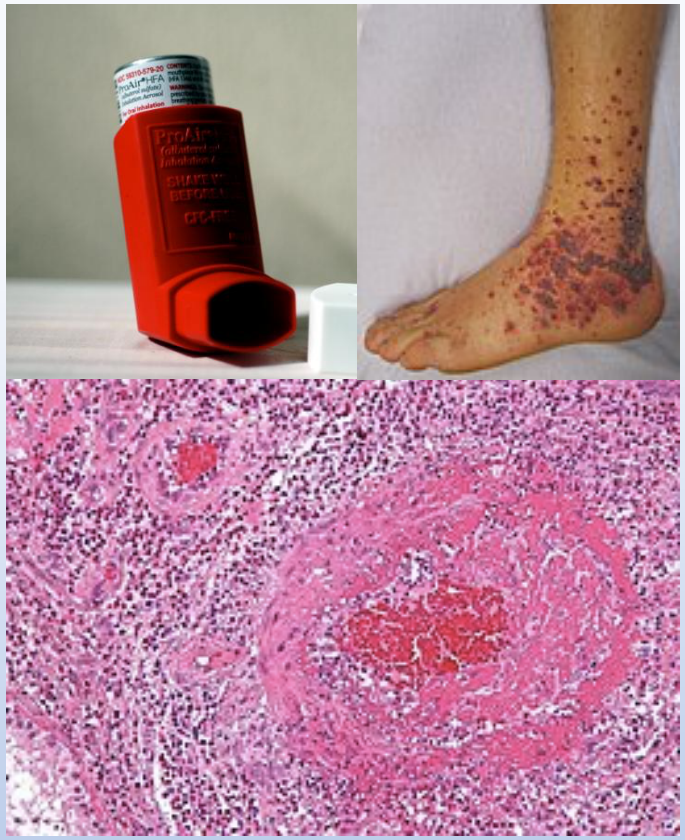
EGPA symptoms can vary greatly

- Key features are asthma, eosinophilia, and vasculitis
- Some present with constitutional symptoms (weight loss, etc)
- While asthma and vasculitis are more common symptoms, EGPA can present in any organ

Phasic Presentations

EGPA symptoms can vary

- Key features are asthma, eosinophilia, and vasculitis but they may or may not appear in that order (or at all)
- Some present with constitutional symptoms (weight loss, etc)
- While asthma and vasculitis are more common symptoms, EGPA can present in any organ



Organs Affected

Lungs

- 90% of EGPA have asthma-like symptoms
- Most asthma will be steroid-dependent
- Rhinitis, sinusitis, and nasal polyps common

Skin

- Rash is common
- Hives, ulcers may also occur

Blood

- Eosinophils > 1200/ml

Heart

- 25% - 30% of EGPA show cardiac involvement
  - Myocarditis
  - Myocardial inflammation
  - Pericarditis
  - Cardiomyopathy

Peripheral nervous system

- Numbness, tingling, weakness
- Mostly hands, feet

Central nervous system

- Stroke symptoms

Furuta et al *Allergology International* 2019
Groh et al. *Eur J Intern Med* 2015

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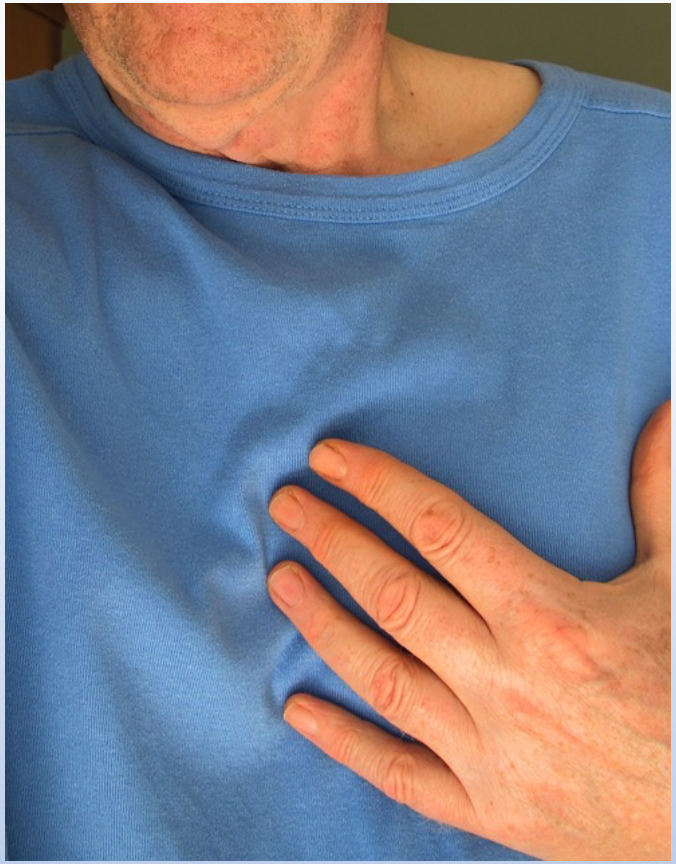


Furuta et al *Allergology International* 2019
Groh et al. *Eur J Intern Med* 2015

Organs Affected

GI tract

- Eosinophilic gastroenteritis and/or mesenteric vasculitis can occur
  - Abdominal pain, nausea, vomiting, diarrhea

Kidneys

- Necrotizing glomerulonephritis
- May lead to hypertension

Other

- ENT symptoms, thromboembolic disease, salivary glands, vasculitis of breasts

Furuta et al *Allergology International* 2019. Chakraborty. *StatPearls* 2019.
Groh et al. *Eur J Intern Med* 2015

ANCA

Anti-neutrophil cytoplasm antibody (ANCA)

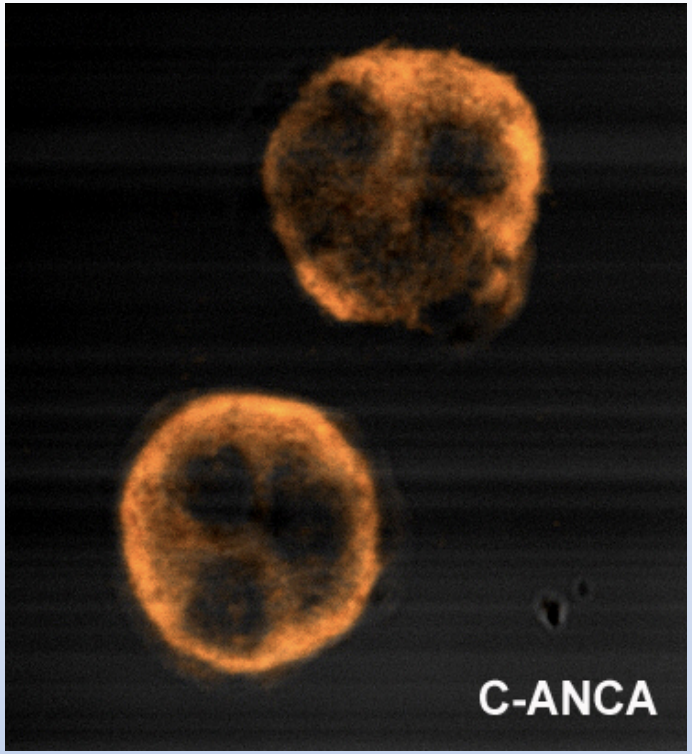
- EGPA is recognised as one form of ANCA-associated vasculitis

ANCA positive EGPA

- Glomerulonephritis and neuropathy more common
- Linked to *HLA-DRB1* gene mutation
- Rituximab response very good

ANCA negative EGPA

- Heart failure more common
- Linked to *IL-5* gene mutation
- Less responsive to Rituximab



Furuta et al *Allergology International* 2019.

Summary

Key takeaways

- Early symptoms make diagnosis difficult
- Multiple organs can be affected
- ANCA a factor