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

Module 1: EGPA Explained

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Case Study

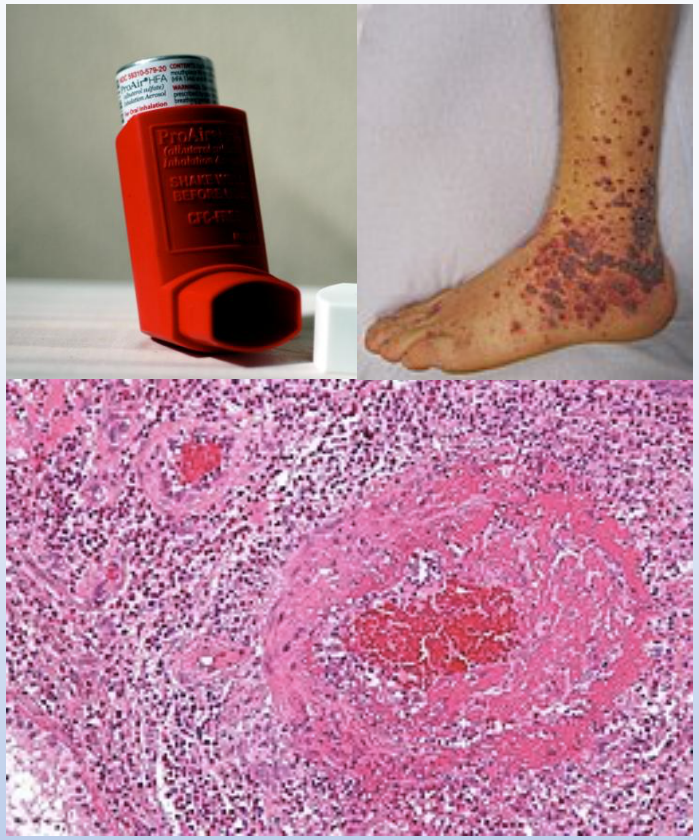
53-yr old woman with shortness of breath

- Makes appt with GP
- Assessment
  - Bilateral pulmonary infiltrate
  - Eosinophil count 1600/ml (21% of total wbc)
- Treated with antibiotic and steroids
  - Symptoms improved

Case Study

Three weeks later

- Shortness of breath, left leg weakness and rash
- Assessment
  - Eosinophil count 28%
  - Biopsy of rash
  - EMG of leg
  - Echocardiogram
- Diagnosed with EGPA



Case Study

Why diagnosed with EGPA

- EGPA is difficult to diagnosis
- When clinical manifestations include asthma, eosinophilia, pulmonary infiltrates and vasculitis, a diagnosis of EGPA is fairly straight forward
- Unfortunately, many cases of EGPA are not that straightforward
- Also, this ‘quick diagnosis’ did not take into account the years the person likely had of incidental rashes, breathing concerns, and unreported cases of numbness

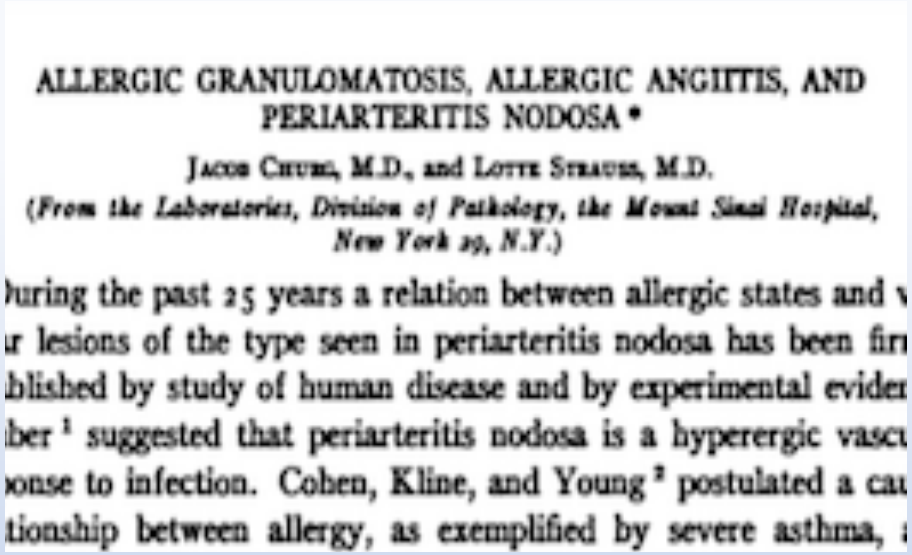
A Quick History Lesson

1951

- Drs Jacob Churg and Lotte Strauss at Mount Sinai Hospital in NYC published a report looking at 13 autopsies.
- Disease named Churg-Strauss Syndrome

2012

- Chapel Hill Consensus Conference
- Renamed eosinophilic granulomatosis with polyangiitis (EGPA)



ALLERGIC GRANULOMATOSIS, ALLERGIC ANGIITIS, AND PERIARTERITIS NODOSA *

JACOB CHURG, M.D., and LOTTE STRAUSS, M.D.

(*From the Laboratories, Division of Pathology, the Mount Sinai Hospital, New York 29, N.Y.*)

uring the past 25 years a relation between allergic states and v
r lesions of the type seen in periarteritis nodosa has been firs
blished by study of human disease and by experimental evider
ber [1] suggested that periarteritis nodosa is a hyperergic vascu
onse to infection. Cohen, Kline, and Young [2] postulated a cau
tionship between allergy, as exemplified by severe asthma,

Churg, Strauss. *Am J Pathol.* 1951.
Jennette et al *Arthritis Rheum.* 2013.

EGPA By the Numbers

Prevalence

- 10-15 cases per one million
- Men and women equally

Age

- Age of symptom onset usually in their 40s
- Age of diagnosis usually in their 50s
- Median duration of asthma onset to diagnosis is 5 – 9 years

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

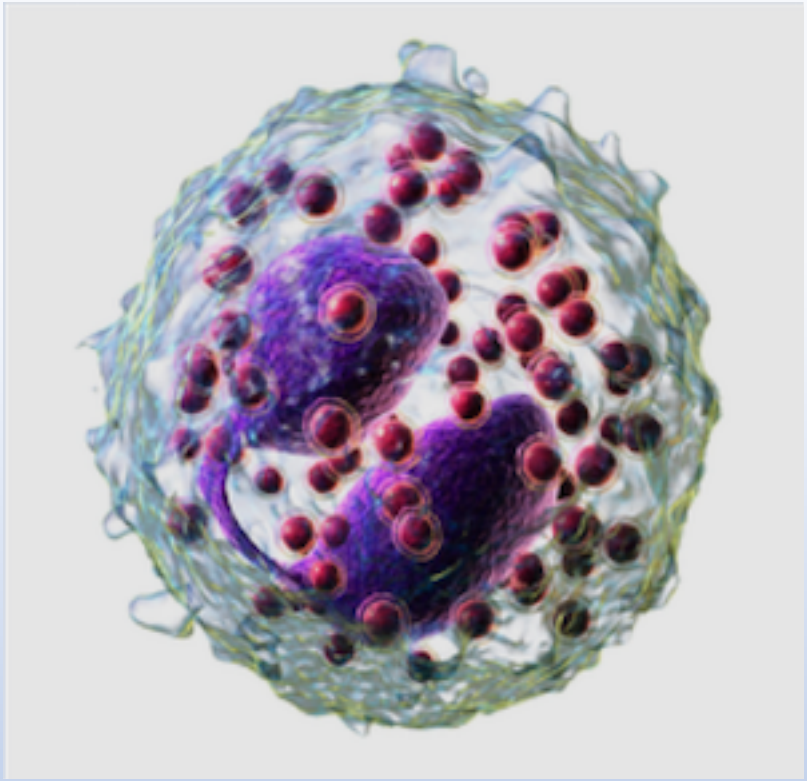
Pathophysiology

Three major pathophysiologic features

- Eosinophil driven (Type 2/TH2/ILC2)
- Neutrophil driven (nontype 2)
- B-cell driven (ANCA)

Nature vs nurture

- Familial component
- Mutations of *IL-10* and *HLA-DRB4* genes linked to EGPA
- Environmental factors and infections may be linked to EGPA



Gioffredi et al *Front Immunol* 2014
Furuta et al *Allergology International* 2019

Pathophysiology

Current understanding

- The cause of EGPA is unknown; however, an allergic mechanism, with tissue directly injured by eosinophils and neutrophil degranulation products, may be involved
- Activation of T lymphocytes seems to help maintain eosinophilic inflammation
- The syndrome occurs in patients who have adult-onset asthma, allergic rhinitis, nasal polyposis, or a combination
- Antineutrophil cytoplasmic autoantibodies (ANCA) are present in about 40% of cases

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

Summary

Key takeaways

- EGPA is difficult to diagnose
- Age of onset usually in a person's 40s. Median time to diagnosis 5 - 9 years
- The cause of EGPA is unknown; however, an allergic mechanism, with tissue directly injured by eosinophils and neutrophil degranulation products, may be involved