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Eosinophilic Myocarditis Secondary to Hypereosinophilic Syndrome vs. Eosinophilic Granulomatosis with Polyangiitis: A Diagnostic Dilemma

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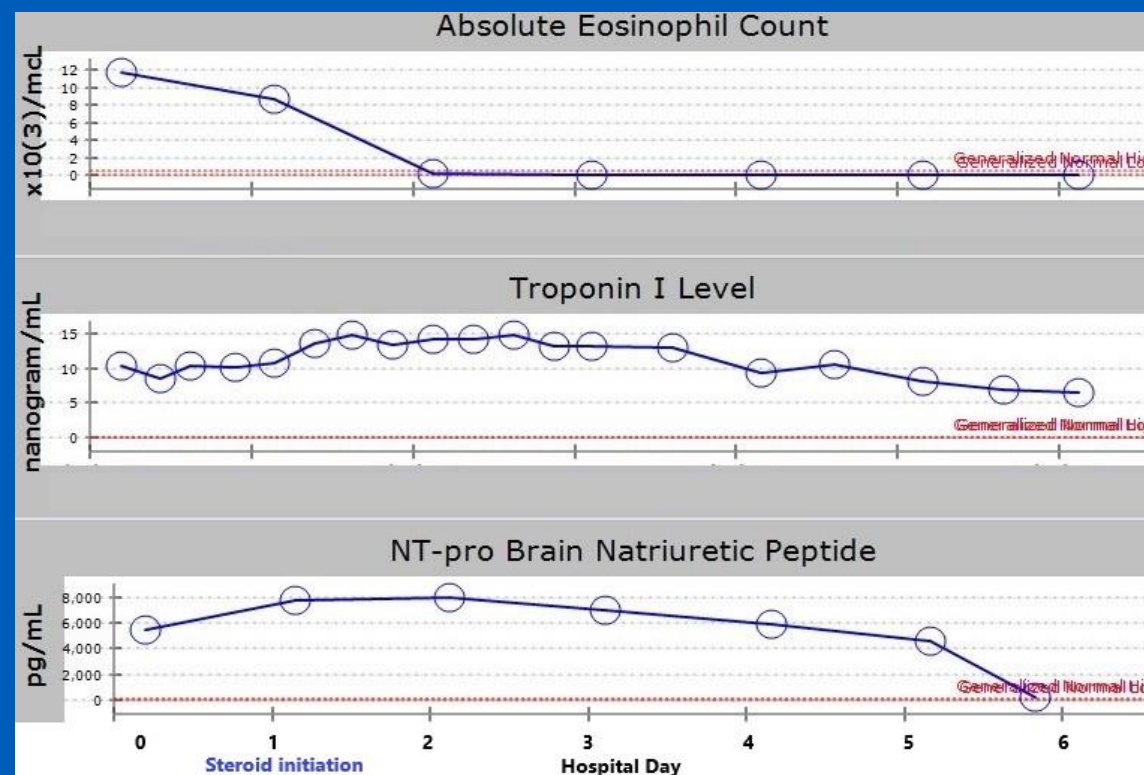
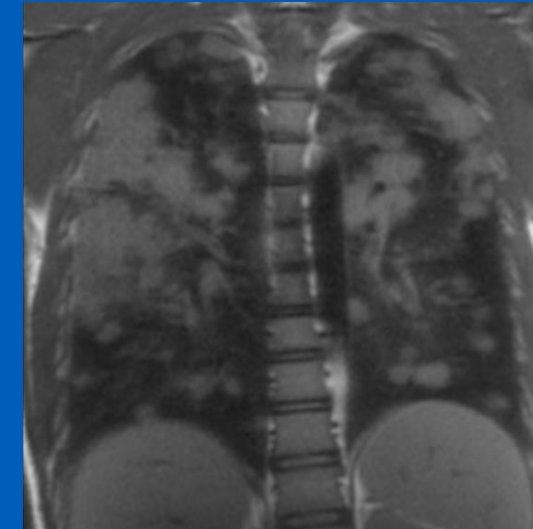
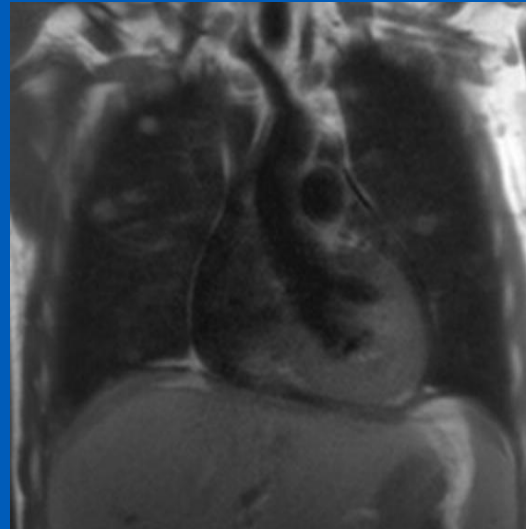
INTRODUCTION

- HES is a rare diagnosis
- Cardiac involvement can lead to irreversible damage and death
- Overlap exists between HES and EGPA, requiring a multi-disciplinary approach

CASE

- 16 y/o male with history of asthma presents with acute onset of chest pain
- Peripheral eosinophilia of $11.7 \times 10^3/\text{mcl}$
- Troponin elevation of 10.4 (ng/mL)
- ECG: ST elevations, multifocal PVCs, and rate dependent BBB
- Cardiac MRI: moderate to severe dysfunction, moderate effusion
- Extensive genetic and infectious work up
- High dose steroids initiated → improvement in eosinophilia, cardiac function, and arrhythmia burden
- Recurrent eosinophilia and effusion → transitioned to therapy with mepolizumab
- Overall improved function

Eosinophilic Myocarditis Secondary to Hypereosinophilic Syndrome vs. Eosinophilic Granulomatosis with Polyangiitis: A Diagnostic Dilemma



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HES

- Hereditary, Primary, Secondary, Unknown
- AEC >1500 or tissue hypereosinophilia x2, 1mo apart for >6mo
- 1. Must be no other cause
- 2. Signs/symptoms of organ involvement
- Abnormal bone marrow biopsy
- JAK2, V617F, c-kit D816V, FIP1L1/PDGFR alpha gene mutations
- Can present with myocarditis

EGPA

1. Asthma
 2. Peripheral eosinophilia (>10% and/or >1500 cells/microL)
 3. Neuropathy
 4. Pulmonary opacities
 5. Paranasal sinus abnormality
 6. Eosinophils on biopsy
- Polyangiitis/surrogates of vasculitis

Conclusion

- HES requires prompt recognition and treatment
- Initial management of HES and EGPA are similar but refractory therapies differ