Neurological Marvels: Unraveling the Enigma of Organ-Specific Hypereosinophilic Syndrome

Sidra Tahir, MD[1]; Giann J Bhatt, DO[1]; Bernard S Shalit, MD[2]; Kyle Koteleba[2]; Mohammed Naveed Hasan [3]

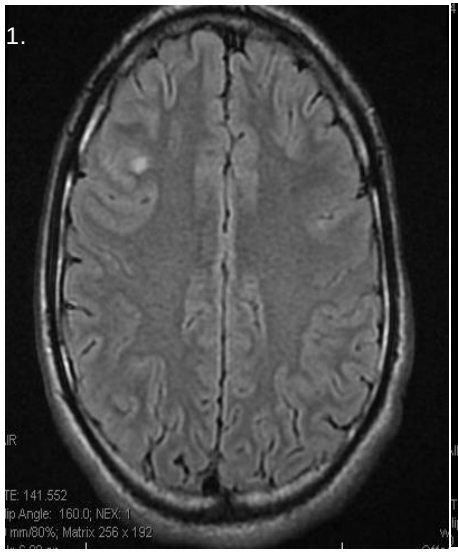
[1]Department of Internal Medicine, University of Pittsburgh Medical Center, Mercy Hospital, Pittsburgh, PA; [2]Department of Rheumatology, University of Pittsburgh Medical Center, Mercy Hospital, Pittsburgh, PA [3]Department of Infectious Diseases University of Pittsburgh Medical Center, Mercy Hospital, Pittsburgh, PA; [4]Department of Neurosurgery, University of Pittsburgh Medical Center, Mercy Hospital, Pittsburgh, PA

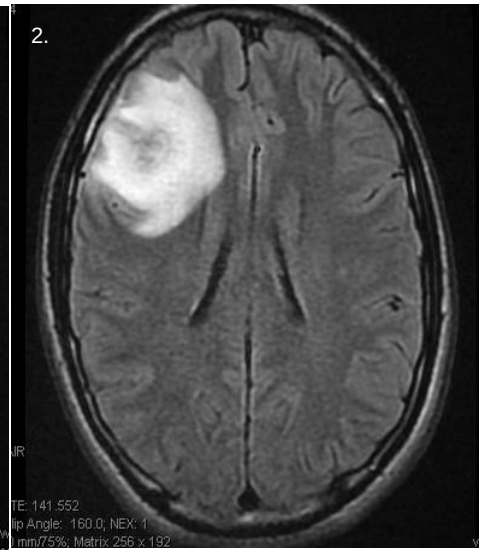
Introduction

- Hypereosinophilic syndrome (HES): a group of disorders characterized by an eosinophil count of >1500 cells/mL or >10% of leukocyte differential for over six months, and/or pathologic confirmation of tissue involvement.
- Typically manifests with organ involvement in the skin, lungs, and gastrointestinal tract
- Primary neurological manifestation of HES, far uncommon, predominantly presents as peripheral neuropathy or cerebral thromboembolism

Case presentation

- 18-year-old male with a history of eosinophilic esophagitis presented with a new-onset seizure preceded by an episode of sinusitis.
- MRI :two small lesions within the right frontal lobe, empirically treated for meningitis. 13.7% eosinophilia.
- CSF studies: mildly increased WBCs and protein.
- Despite antibiotics, serial MRIs demonstrated progression of the lesion
- Biopsy: perivascular lymphocytes with focally prominent eosinophils.
- Positive only for ESR of 53 and a weakly positive ANA, with otherwise negative Rheumatologic workup inc ANCA.
- Pulse dose steroids x5 days followed by taper x 3m.Outpatient MRI almost complete resolution of mass.





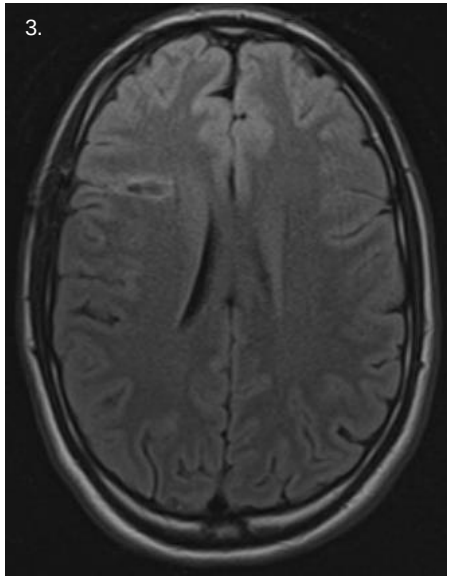


Figure1:
MRI on presentation post seizure

Figure 2 :
MRI post 12 days of Antibiotics

Figure 3:
Outpatient MRI 5 months post discharge

Discussion

- Pathophysiology of HES, two mechanisms proposed; impairment of eosinophil apoptosis by increased levels of IL-5 and clonal proliferation of eosinophils during hematopoiesis.
- The absence of ANCA in this case does not rule out EGPA as ANCA positivity is present in only 40-60% of cases. Debate persists whether EGPA (which tends to be more organ specific) is a subtype of HES or its own independent syndrome
- Hypothesis for brain lesion: eosinophil-induced endothelial damage, facilitating eosinophilic transmigration into the CNS, potentially triggered by the patient's preceding sinus infection.
- A unique dimension due to primary neurologic involvement in HES which challenges the presentations documented in previous case reports, which usually involve primarily peripheral neuropathy, encephalopathy[1], or cerebral thromboemboli.

Learning Point

Organ-Specific HES, as evidenced by eosinophilic infiltrate on brain biopsy of our patient, is a distinctive subtype of HES that is characterized by single organ involvement that may or may not have eosinophilia > 1500 cells.

[1] *Moore PM, Harley JB, Fauci AS. Neurologic dysfunction in the idiopathic hypereosinophilic syndrome. Ann Intern Med. 1985 Jan;102(1):109-14. doi: 10.7326/0003-4819-102-1-109. PMID: 2981493.*

tahirs3@upmc.edu