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Eosinophilic granulomatosis with polyangiitis: A unique case of secondary medium vessel vasculitis and temporal arteritis

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Description

A 60-year-old male presented with a 1-month history of ocular photosensitivity, pruritus, infraorbital swelling, and acute binocular vision loss. He was admitted for evaluation of anterior optic neuropathy and temporal arteritis. Neuro-Ophthalmology recommended 1 g methylprednisolone, leading to significant improvement in right eye vision but persistent left eye deficits. Laboratory tests showed elevated CRP (185.9 mg/L), ESR (128 mm/hr), rheumatoid factor (61 IU/mL), and eosinophilia (absolute 3.06 x 10^9/L). CT Head and MRI revealed ethmoid cell opacification, soft tissue stranding, and diffuse paranasal sinus mucosal thickening. CT chest revealed pulmonary nodules (Figure 1A; blue arrows denoting lung nodules), ground-glass opacities, and cervical lymphadenopathy. Temporal artery ultrasound (patient declined biopsy) showed decreased compressibility, and a halo sign around the left temporal artery, concerning for giant cell arteritis (Figure 1B and 1C) [1]. He was started on Tocilizumab. During hospitalization, he developed a purpuric lower extremity rash (Figure 1D and 1E; orange arrows denoting rash)—biopsy confirmed medium vessel vasculitis (Figure 1F). He was started on cyclophosphamide induction therapy and a prednisone taper, on which he had symptomatic and radiographic improvement post discharge.

(Figure 1G), though eosinophilia persisted (absolute 1.02 x 10^9/L, previously 1.08 x 10^9/L). Work-up for hyper-eosinophilic malignancy (Leukemia/lymphoma immunophenotyping, T-cell rearrangement, chronic eosinophilia panel) was negative. Following the 3rd dose of cyclophosphamide, he experienced symptom relapse and was started on mepolizumab with improvement. Based on the ACR/EULAR criteria, he scored 10 points due to eosinophilia ≥1 × 10^9/L, obstructive lung disease of pulmonary function testing, and extravascular eosinophilic predominant inflammation seen on his skin biopsy [2].

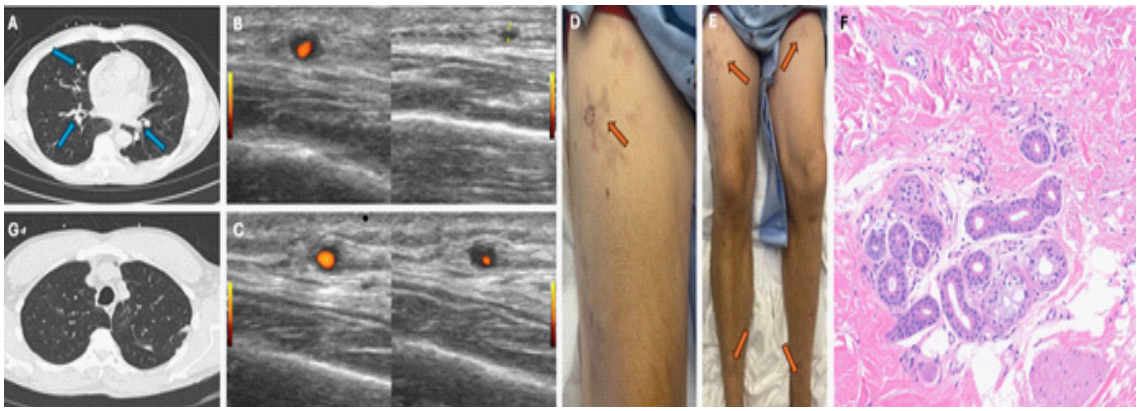

Figure 1: Clinical image.

Ethics: Written informed consent was obtained for publication. IRB approval/waiving was not required per Mayo Clinic policy given that the study is based on a single patient.

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