Paracentral Acute Middle Maculopathy Associated with Giant Cell Arteritis

M. Zia Siddiqui, MD; Athanasios Papakostas, MD

Introduction:

An 86-year-old female patient was referred to our retina clinic for subacute vision loss in the left eye over 2 weeks. Her past medical history was significant for hypertension, hypothyroidism, chronic obstructive pulmonary disease, and coronary artery disease. Her ocular history was significant for early dry age-related macular degeneration and pseudophakia of both eyes.

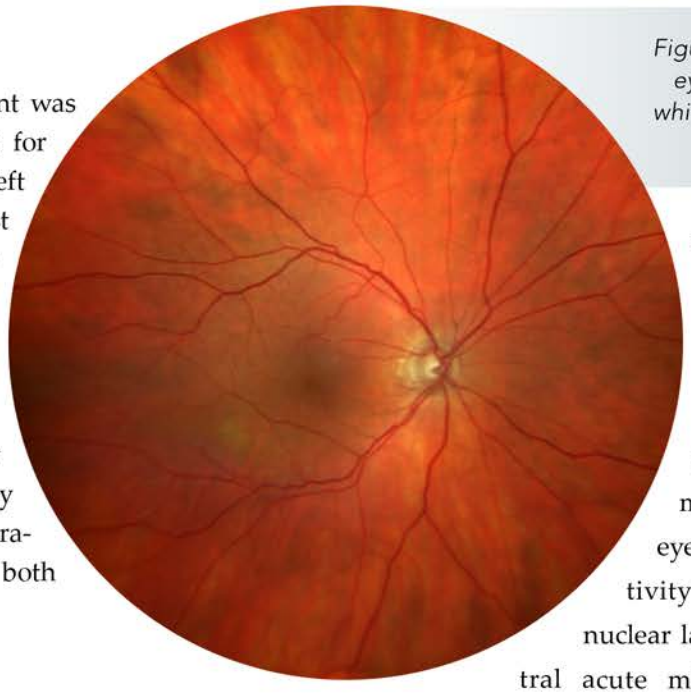
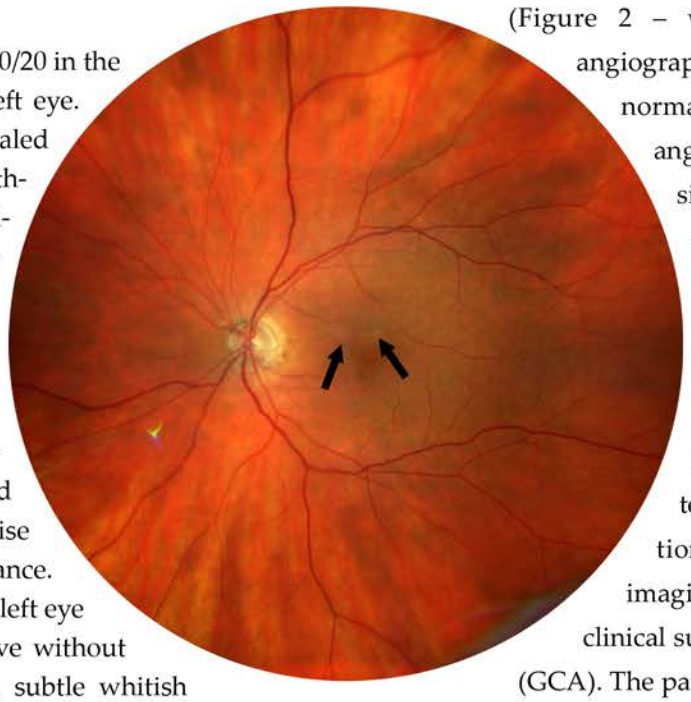


Figure 1: Fundus photos of both eyes. Arrows indicate subtle whitish parafoveal abnormalities in the left eye.

Examination:

On exam, visual acuity was 20/20 in the right eye and 20/80 in the left eye. The pupil examination revealed reactive and equal pupils without a relative afferent pupillary defect. Intraocular pressures were 16mmHg and 14mmgHg. The anterior segment exam was unremarkable for both eyes. Posterior segment examination of the right eye revealed macular drusen and otherwise normal funduscopic appearance. The posterior segment of the left eye revealed a normal optic nerve without hyperemia, macular drusen, subtle whitish parafoveal appearance (Figure 1 – black arrows), and normal peripheral examination.



OCT macula of the right eye was consistent with macular drusen and otherwise normal. OCT macula of the left eye showed areas of hyperreflectivity at the level of the inner nuclear layer consistent with paracentral acute middle maculopathy lesions (Figure 2 – white arrows). Fluorescein angiography of the right eye revealed normal filling times. Fluorescein angiography of the left eye was significant for areas of delayed choroidal filling (Figure 3). On external exam, the patient had a prominent left superficial temporal artery. On further questioning, the patient reported temporal tenderness and jaw claudication. Given the clinical and imaging findings, there was high clinical suspicion for giant cell arteritis (GCA). The patient was emergently referred to the emergency room for laboratory work-up and

high-dose systemic steroids. Laboratory values showed a normal erythrocyte sedimentation rate (ESR) at 2 mm/hr, and the C-reactive Protein (CRP) was markedly elevated at 97 mg/L. The patient underwent a temporal artery biopsy, which revealed giant cell arteritis. She continued high-dose intravenous systemic steroids and was discharged with an oral prednisone taper and rheumatology referral. The patient's vision remained stable with subjective improvement at subsequent follow-up.

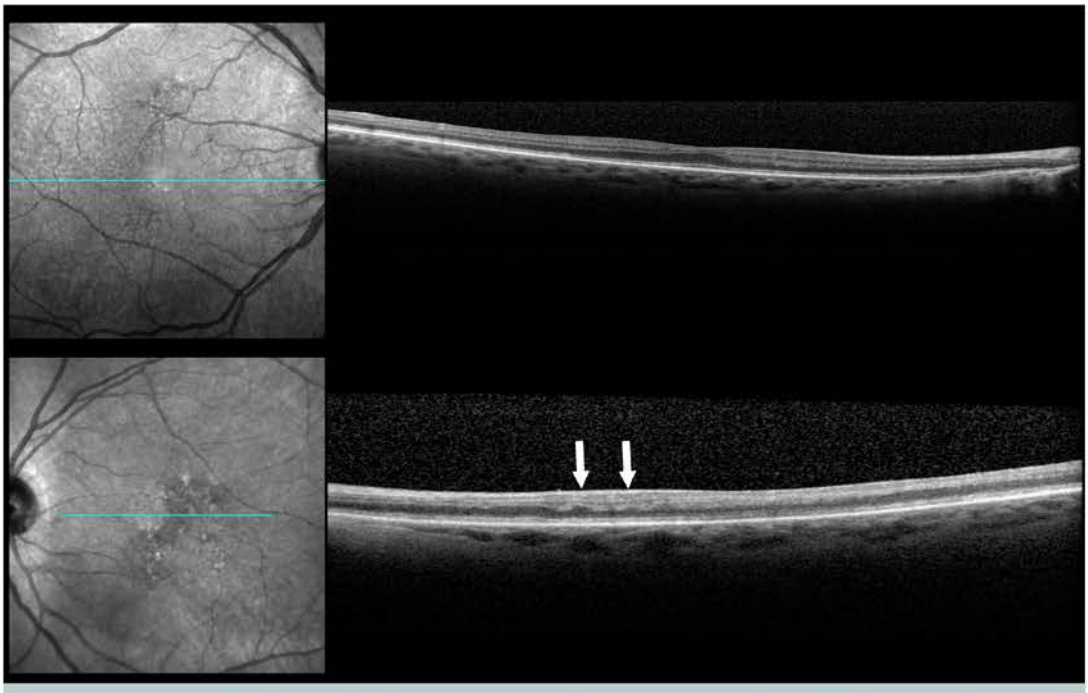


Figure 2: OCT of the right eye was significant for macular drusen and RPE changes. OCT of the left eye showed areas of hyperreflective bands at the level of the inner nuclear layer consistent with paracentral acute middle maculopathy lesions (white arrows).

Discussion:

Giant cell arteritis (GCA) is an immune-mediated primary systemic granulomatous vasculitis that can lead to profound vision loss in one or both eyes.[1] Early diagnosis is essential to prevent permanent vision loss and systemic complications. Ocular involvement varies widely, from 20% to 70%, so a high degree of suspicion is necessary.[1] While paracentral acute middle maculopathy (PAMM) may manifest independently, it can also arise from systemic vascular disorders as PAMM lesions are indicative of localized retinal ischemia in the deep capillary plexus.[2] Recent literature has supported that isolated paracentral acute middle maculopathy (PAMM) can be an early sign of GCA where vision loss may be less profound.[3] In our specific case, spectral domain OCT imaging had findings of hyper-reflectivity

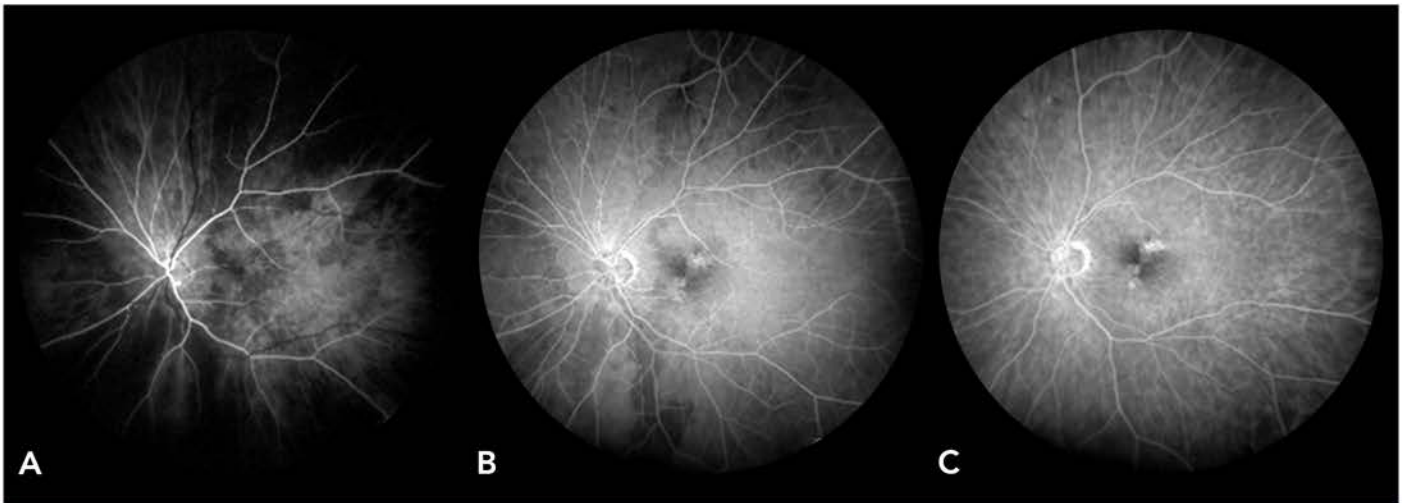


Figure 3: Fluorescein angiography of the left eye - (A) Early phase (B) Mid phase (C) Late phase. FA was significant for delayed choroidal filling.

in the inner nuclear layer and choroidal hypoperfusion on fluorescein angiography.[1,2] Given the imaging findings and positive review of the system, there was a high suspicion of giant cell arteritis. Management for giant cell arteritis includes emergent referral for work-up and high-dose systemic steroids. Additionally, we believe that PAMM lesions in patients with a positive review of the system should be emergently investigated for GCA and started on high-dose systemic steroids.

References:

1. Hayreh S. Giant cell arteritis: Its ophthalmic manifestations, Indian J Ophthalmology 2021; 69(2): 227-235.

2. Abtahi SH, Nourinia R, et al. Retinal ischemic cascade: New insights into the pathophysiology and imaging findings, Survey of Ophthalmology 2023; e380-e387.

3. Broyles H, Chacko J, et al. Paracentral Acute Middle Maculopathy as the Initial Presentation of Giant Cell Arteritis, Journal of Neuro-Ophthalmology 2021; 41: e157-e159.

THE RETINA INSTITUTE

2201 Brentwood Blvd.
St. Louis, MO 63144

(314) 367-1181
tri-stl.com

Kevin J. Blinder, MD
Barton L. Blackorby, MD
Sabin Dang, MD
Alia K. Durrani, MD
Nicholas E. Engelbrecht, MD

Avinash V. Honasoge, MD, PhD
Nicholas A. Iafe, MD
Thomas K. Krummenacher, MD
Athanasios Papakostas, MD
Bradley T. Smith, MD