**CLINICAL VIGNETTE**

# Navigating Diagnostic Challenges in Giant Cell Arteritis

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***Case Presentation***

A 64-year-old female with a history of pre-diabetes, hypertension, and hyperlipidemia presented for left temporal headache with associated jaw claudication symptoms for two weeks. Pain increased when she lay on her left side and was associated with left eye pain without tearing, vision loss, or vision changes. She denied recent weight loss, fevers, chills, weakness or neuropathic symptoms in her extremities, no new rashes, joint pain or swelling was noted. She denied family history of autoimmune disease, prior heart attack, or stroke. Her medications included rosuvastatin and metoprolol.

On physical exam, her pupils were equal and reactive to light, no lymphadenopathy was palpated. She noted tenderness to palpation of the left temporal region and jaw with an intact temporal pulse palpated bilaterally. She did not have any observed rashes, nor joint tenderness or synovitis. Her cranial nerves were intact and she had no nuchal rigidity. Her pulmonary, cardiovascular and abdominal exams were normal.

She underwent computed tomogram (CT) head and facial imaging which was normal. Her complete blood count (CBC) and basic metabolic panel (BMP) were normal. Her erythrocyte sedimentation rate (ESR) resulted at 38 (N<30 mm/hr) and her C-reactive protein (CRP) resulted 25.6 (N<0.3 mg/dL). Neurology and rheumatology were consulted. Magnetic resonance imaging (MRI) brain was obtained and was normal. Prednisone initiation in the emergency department was recommended by neurology. However, further doses were held after the patient was evaluated by the rheumatology service and noted to have minimal headache symptoms, to increase yield from temporal artery biopsy. Vascular surgery was consulted but due to logistical reasons, biopsy was not performed until 72 hours after initial consultation. During this period the patient reported increased headache severity and oral prednisone 40mg was started. Pathology subsequently confirmed the diagnosis (see Figures 1 and 2) and she was started on high dose prednisone (1mg/kg) with plan for prolonged outpatient taper after dis-charge to home.

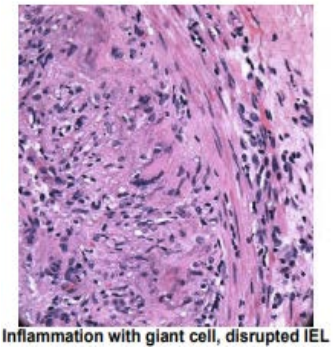
***Discussion***

Giant cell arteritis (GCA) is a chronic inflammatory condition, primarily affecting medium to large arteries. The diagnosis of this condition relies on a combination of clinical findings, laboratory results, imaging and biopsy. Timely diagnosis is crucial in preventing the feared outcome of vision loss, but accurate diagnosis need also be scrutinized as the first line treatment of prolonged high dose steroids has the potential for major health implications for patients. We will highlight the clinical complexities of diagnosis and treatment initiation of this less common disease state.[1-3]

This case provides an overview of the important points in history taking and examination and highlights the complexity of initial management in patients presenting with high suspicion for GCA. The most feared complication of GCA is vision loss, but the timing of diagnostics and symptom management create a complex balancing act. In our patient, the 2022 ACR/ EULAR classification criteria were used with a moderate to high suspicion for GCA.[4] Temporal artery biopsy was delayed due to logistical reasons with vascular surgery. Treatment was initiated due to persistent symptoms. Biopsy remains the gold standard for diagnosis but is subject to sampling yield error and medication effects. Preferred imaging modality remains uncertain with ultrasound, positron emission tomography (PET)/CT, MRI all without clear advantage over each other.[5]

Our patient was treated with high dose steroids which remains first line treatment. High dose steroids often continue for a month before slow tapering over the subsequent months to year.[6] Of note, tocilizumab (an anti-IL6 receptor antibody involved within the inflammatory process) was approved by the FDA in 2017 for GCA treatment. Studies report increased remission rates and reduced steroid use over 1 year when used in combination. However, further studies are needed to determine whether monotherapy with tocilizumab remain more effective than steroids alone and clinical practice has yet to shift from high dose steroid administration. However, tocilizumab monotherapy is an alternative option to consider in patients unable to tolerate long term high dose steroids.[7]

***Figures***



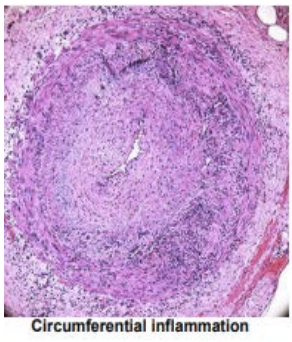




Figure 1. Inflammation with giant cell and disrupted internal elastic lamina of the temporal artery.
Figure 2. Circumferential inflammation with the presence of lymphocytes, plasma cells and histiocytes.

**REFERENCES**


1. **Buttgereit F, Dejaco C, Matteson EL, Dasgupta B**. Polymyalgia Rheumatica and Giant Cell Arteritis: A Systematic Review. *JAMA*. 2016 Jun 14;315(22):2442-58. doi: 10.1001/jama.2016.5444. PMID: 27299619.
2. **Lyons HS, Quick V, Sinclair AJ, Nagaraju S, Mollan SP**. A new era for giant cell arteritis. *Eye (Lond)*. 2020 Jun;34(6):1013-1026. doi: 10.1038/s41433-019-0608-7. Epub 2019 Oct 3. PMID: 31582795; PMCID: PMC7253415.
3. **Koster MJ, Matteson EL, Warrington KJ**. Large-vessel giant cell arteritis: diagnosis, monitoring and management. *Rheumatology (Oxford)*. 2018 Feb 1;57(suppl_2):ii32-ii42. doi: 10.1093/rheumatology/kex424. PMID: 29982778.
4. **Ponte C, Grayson PC, Robson JC, Suppiah R, Gribbons KB, Judge A, Craven A, Khalid S, Hutchings A, Watts RA, Merkel PA, Luqmani RA; DCVAS Study Group**. 2022 American College of Rheumatology/ EULAR Classification Criteria for Giant Cell Arteritis. *Arthritis Rheumatol*. 2022 Dec;74(12):1881-1889. doi: 10.1002/art.42325. Epub 2022 Nov 8. PMID: 36350123.
5. **Moreel L, Betrains A, Doumen M, Molenberghs G, Vanderschueren S, Blockmans D**. Diagnostic yield of combined cranial and large vessel PET/CT, ultrasound and MRI in giant cell arteritis: A systematic review and meta-analysis. *Autoimmun Rev*. 2023 Jul;22(7):103355. doi: 10.1016/j.autrev.2023.103355. Epub 2023 May 3. PMID: 37146926.
6. **Allison MC, Gallagher PJ**. Temporal artery biopsy and corticosteroid treatment. *Ann Rheum Dis*. 1984 Jun;43(3): 416-7. doi: 10.1136/ard.43.3.416. PMID: 6742903; PMCID: PMC1001361.
7. **Stone JH, Tuckwell K, Dimonaco S, Klearman M, Aringer M, Blockmans D, Brouwer E, Cid MC, Dasgupta B, Rech J, Salvarani C, Schett G, Schulze-Koops H, Spiera R, Unizony SH, Collinson N**. Trial of Tocilizumab in Giant-Cell Arteritis. *N Engl J Med*. 2017 Jul 27;377(4):317-328. doi: 10.1056/NEJMoa1613849. PMID: 28745999.