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Enhancement within the orbits and superficial temporal artery: MRI clues to the diagnosis of giant cell arteritis

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ABSTRACT

Purpose: Giant cell arteritis can have protean manifestations, many of them non-specific. We describe a patient whose initial presentation was most consistent with brainstem stroke, but who was ultimately diagnosed with and treated for giant cell arteritis.

Observations: A 76-year-old woman presented with abrupt onset diplopia, headache, nausea and vomiting. On presentation to the emergency department, she was diagnosed with a brainstem stroke and treated with tissue plasminogen activator. When her symptoms did not remit, closer inspection of her imaging revealed enhancement within the orbits and enhancement of her superficial and posterior temporal arteries. These findings led to the consideration of giant cell arteritis. Her signs and symptoms resolved with steroid treatment and a temporal artery biopsy was positive.

Conclusions and importance: This case highlights the emerging role of MRI and other imaging techniques in improving our ability to non-invasively diagnose giant cell arteritis.

1. Introduction

Giant cell arteritis can have protean manifestations, many of them non-specific. We describe a patient whose initial presentation was most consistent with brainstem stroke, but who was ultimately diagnosed with and treated for giant cell arteritis.

2. Case report

A 76-year-old Caucasian woman awakened with binocular oblique diplopia, severe headache, nausea and vomiting. On arrival in the emergency department, an acute stroke code was initiated. A CT scan of the head was normal and tissue plasminogen activator was administered for treatment of suspected brainstem stroke. She was admitted to the neuro intensive care unit.

The patient, a retired physician, had a past ocular history of mild age-related cataracts and macular degeneration. Her past medical history was remarkable for Merkel cell carcinoma (in remission), deep venous thrombosis and pulmonary embolism following shoulder surgery.

Best-corrected visual acuity was 20/25 in the right eye and 20/20 in the left eye and intraocular pressure was normal in both eyes. Pupils were normal. Although extraocular motility was grossly normal, alternate cover testing revealed a comitant right hypertropia, felt to be consistent with a skew deviation. She had no imbalance, dysmetria or nystagmus. Anterior segments were quiet with mild nuclear sclerotic cataracts and dilated fundus examination was normal aside from hard drusen in the macula.

The following day, her nausea and vomiting were medically controlled, but her headache and double vision had not improved. Erythrocyte sedimentation rate was 16 (normal <30 mm/hr) and C-reactive protein was 7.4 (normal 0.0–0.8 mg/dL). An MRI without contrast did not reveal a stroke. Specifically, there was no diffusion restriction evident in the brainstem. Two days later, with her diplopia and headache only minimally improved, a high-resolution MRI of the orbits

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was completed. High-resolution images of the brain with post-contrast black-blood vessel wall images were also obtained. The MRI of the orbits was remarkable for enhancement of the optic nerve sheaths and stranding of orbital fat in each orbit (Fig. 1). Because enhancement of orbital structures has been described in patients with giant cell arteritis (GCA), this diagnosis was considered more carefully. On suspicion of GCA, 1 g of intravenous methylprednisolone was administered with a marked improvement in her headache and diplopia. A temporal artery biopsy revealed a thinned intima and absent internal elastic lamina. There was an intense inflammatory reaction with epithelioid cells and multinucleated giant cells. GCA was diagnosed based on these pathologic findings. She received a total of three infusions of intravenous methylprednisolone (1 g each) and was discharged on 60 mg oral prednisone with complete resolution of her headache and diplopia. Retrospective evaluation of the MRI brain with contrast revealed enhancement of the left superficial and posterior temporal arteries (Fig. 2).

She returned to her out-of-state home where she was evaluated by a rheumatologist. She was started on injections of tocilizumab as her prednisone dose was tapered without recurrence of symptoms.

3. Discussion and conclusions

The constellation of skew deviation, nausea and vomiting is an atypical presentation of GCA. This patient's diplopia was most likely due to orbital involvement by GCA, with ischemia of one or several extraocular muscles. We considered the possibility of a brainstem stroke; however, this was deemed much less likely due to the absence of associated neurological deficits and the lack of corresponding findings on imaging. Ischemia to an ocular motor nerve was also considered, but was not supported by the relative comitance of the deviation. Given the normal sedimentation rate, GCA was not a primary diagnosis until enhancement of the optic nerve sheaths and stranding of orbital fat was noted on MRI. Although non-specific, enhancement of orbital structures has been described in up to 32 % of patients with GCA independent of the presence of orbital symptoms.[1] For this reason, the diagnosis of GCA should be considered if orbital inflammation is detected on MRI.

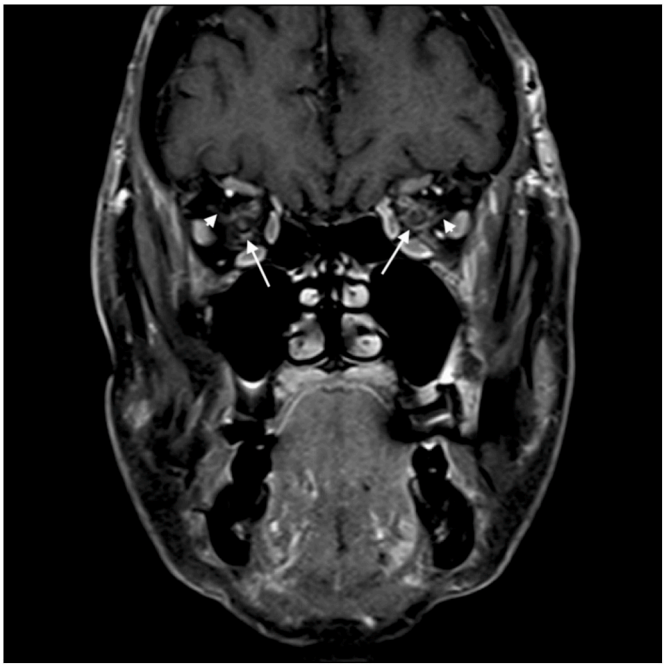

Fig. 1. Coronal T1-weighted, gadolinium enhanced, fat-saturated image of the orbits shows bilateral perineural enhancement of the optic nerve sheaths (arrows) and stranding of orbital fat (arrowheads). Image obtained three days after presentation.

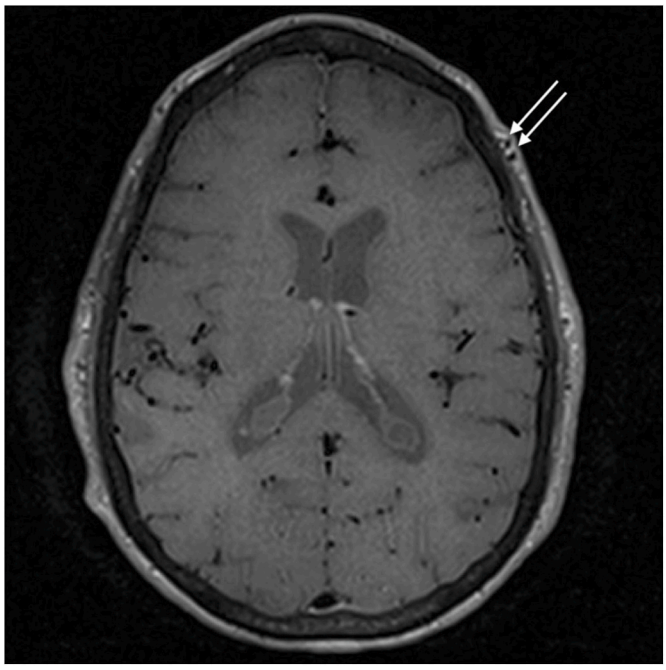

Fig. 2. Axial T1-weighted post-contrast image of the head demonstrates enhancement of the left superficial (top arrow) and posterior temporal arteries (bottom arrow). Image obtained one day after presentation. This study was protocoled using delay alternating with nutation for tailored excitation ("DANTE"). The DANTE sequence can highlight enhancement in vessel walls.

Imaging protocols developed to highlight the status of the head and neck vessel walls have shown diagnostic utility in the setting of giant cell arteritis.[2–4] Vessel wall imaging has proven to be both sensitive and specific and normal vessel wall imaging may obviate the need for superficial temporal artery biopsy when clinical suspicion is low. The negative predictive value of normal vessel wall imaging has been reported to be over 98 %.[4] In this same study, the sensitivity of MRI for GCA was 93.6 %, but there were a substantial number of false positives and the positive predictive value dropped to 48.3 %. To best image the scalp arteries for evidence of vasculitis, specific MRI protocols are required and are described in detail in these reports. It should be noted that positive findings on MRI scanning are strongly affected by corticosteroid treatment and if MRI results are to be used for clinical decision making, the MRI should take place no later than the first few days of corticosteroid treatment.[5]

In a systematic review, Moreel et al.[6] found that combining an ultrasound of the cranial vessels and large vessels had an average sensitivity of 86 % (range = 76–92 %) and an average specificity of 96 % (range = 92–98 %) for detecting GCA. Combining a PET/CT of both the cranial and large vessels yielded an average sensitivity of 82 % (range = 61–93 %) and an average specificity of 79 % (range = 60–90 %) for diagnosing GCA.

Rhee et al.[7] described how MRI scans of the orbits and cranial vessel walls, in combination with an ophthalmic examination, can markedly improve the sensitivity for diagnosing GCA. This manuscript highlights the usefulness of high-resolution vessel wall MRI protocols in maximizing sensitivity and empasizes that the MRI findings rapidly resolve with corticosteroid treatment. This last finding supports the hypothesis that the observed MRI abnormalities represent vessel wall inflammation. The Rhee article features anatomic diagrams and representative images for physicians and researchers who want to better understand the utility of these emerging MRI techniques.

Temporal artery biopsy remains the gold standard for diagnosing GCA. As our ability to image inflammation in the head and neck with

ultrasound, PET/CT and MRI improves, we may be able to rely more confidently on these non-invasive tests.

CRediT authorship contribution statement

George Sanchez: Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Theresa M. Long:** Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Judith E.A. Warner:** Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Lee S. Chung:** Writing – review & editing, Data curation. **Edward Patrick Quigley:** Writing – review & editing, Formal analysis, Data curation. **Kathleen B. Digre:** Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Meagan D. Seay:** Writing – review & editing, Formal analysis, Data curation, Conceptualization. **Alison V. Crum:** Writing – review & editing, Data curation, Conceptualization. **Bradley J. Katz:** Writing – review & editing, Writing – original draft, Supervision, Project administration, Formal analysis, Data curation, Conceptualization.

Patient consent

The patient described in this case report provided verbal consent to publish this report. Written consent was not obtained and this manuscript contains no personal identifying information.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

1. Guggenberger KV, Vogt ML, Song JW, et al. Intraorbital findings in giant cell arteritis on black blood MRI. *Eur Radiol*. 2023 Apr;33(4):2529–2535. https://doi.org/10.1007/s00330-022-09256-7. Epub 2022 Nov 17. PMID: 36394601; PMCID: PMC10017783.
2. Bley TA, Uhl M, Carew J, et al. Diagnostic value of high-resolution MR imaging in giant cell arteritis. *AJNR Am J Neuroradiol*. 2007 Oct;28(9):1722–1727. https://doi.org/10.3174/ajnr.A0638. Epub 2007 Sep 20. PMID: 17885247; PMCID: PMC8134183.
3. Klink T, Geiger J, Both M, et al. Giant cell arteritis: diagnostic accuracy of MR imaging of superficial cranial arteries in initial diagnosis-results from a multicenter trial. *Radiology*. 2014 Dec;273(3):844–852. https://doi.org/10.1148/radiol.14140056. Epub 2014 Aug 6. PMID: 25102371.
4. Rhéaume M, Rebello R, Pagnoux C, et al. High-resolution magnetic resonance imaging of scalp arteries for the diagnosis of giant cell arteritis: results of a prospective cohort study. *Arthritis Rheumatol*. 2017 Jan;69(1):161–168. https://doi.org/10.1002/art.39824. PMID: 27483045.
5. Hauenstein C, Reinhard M, Geiger J, et al. Effects of early corticosteroid treatment on magnetic resonance imaging and ultrasonography findings in giant cell arteritis. *Rheumatology*. 2012 Nov;51(11):1999–2003. https://doi.org/10.1093/rheumatology/kes153. Epub 2012 Jul 6. PMID: 22772317.
6. 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. https://doi.org/10.1016/j.autrev.2023.103355. Epub 2023 May 3. PMID: 37146926.
7. Rhee RL, Rebello R, Tamhankar MA, et al. Combined orbital and cranial vessel wall magnetic resonance imaging for the assessment of disease activity in giant cell arteritis. *ACR Open Rheumatol*. 2024 Jan 24. https://doi.org/10.1002/acr2.11649. Epub ahead of print. PMID: 38265177.